VOLUME LVI OCTOBER, 1946 NUMBER 10

THE LARYNGOSCOPE

FOUNDED IN 1896
BY
MAX A. GOLDSTEIN, M. D.

PUBLISHED BY
THE LARYNGOSCOPE
640 SOUTH KINGSHIGHWAY ST. LOUIS (10), MO., U. S. A.

NOTICE TO CONTRIBUTORS

THE LARYNGOSCOPE reserves the right of exclusive publication of all articles submitted. This does not preclude their publication in Transactions of the various Societies.

Manuscripts should be typewritten, double spaced, on one side of paper only and with sufficient margins to allow for corrections.

References should be complete: author's surname, initials, title of article, journal, volume, page, month, year.

Six illustrations will be furnished for each article without cost to author. Authors will please limit illustrations to six or assume the expense of additional illustrations.

Proofs will be submitted to authors for corrections. If these are not returned, articles will be published as corrected in this office.

Reprints will be furnished at the following prices:

WITHOUT COVER

	250 Copies	500 Copies	1000 Copies	2000 Copies
Four Pages	$ 7.25	$ 8.75	$11.75	$17.50
Eight Pages	15.00	19.00	25.50	38.00
Twelve Pages	22.50	28.50	40.50	64.50
Sixteen Pages	29.00	35.50	47.50	70.00
Twenty Pages	35.00	43.00	59.00	88.00
Twenty-four Pages	42.50	51.50	69.50	104.50
Thirty-two Pages	55.00	66.00	88.00	129.00

WITH COVER

Four Pages	$13.25	$17.00	$24.50	$39.00
Eight Pages	21.00	27.25	38.25	59.50
Twelve Pages	28.50	36.75	53.25	86.00
Sixteen Pages	35.00	43.75	60.25	91.50
Twenty Pages	41.00	51.25	71.75	109.50
Twenty-four Pages	48.50	59.75	82.25	126.00
Thirty-two Pages	61.00	74.25	110.75	150.50

Express charges to be paid by consignee.

THE
LARYNGOSCOPE.

VOL. LVI OCTOBER, 1946. No. 10

DEVELOPMENT OF THE AUDITORY OSSICLES.*†

BARRY J. ANSON, Ph.D. (Med. Sci.) (by invitation),
Chicago, Ill.

INTRODUCTION.

In an examination of more than 250 series of sections, chiefly in Dr. Theodore H. Bast's collection at the University of Wisconsin and in the otological collection at Northwestern University Medical School, the development and the normal adult morphology of the auditory ossicles in man have been studied — from the stage of the 17 mm. embryo to that of the aged adult.

From several articles already published by the present author and his associates,‡ and from material for journal articles now in preparation, crucial observations have been selected for this brief report.

GENERAL CONSIDERATIONS.

In aquatic and amphibious vertebrates, the skeletal elements which, in higher forms become transformed into ossicles, functioned as branchial supports for the respiratory mechanism. These elements are made over to serve as links

*Read at the Seventy-eighth Annual Meeting of the American Otological Society, Inc., Chicago, May 31, 1946.

Investigation conducted under the auspices of the Central Bureau of Research of the American Otological Society.

†Contribution No. 463 from the Department of Anatomy, Northwestern University Medical School.

‡Arch. Otolaryngol., 28:5:676-697, 1938; 29:6:939-973, 1939; 36:6:891-925, 1942. Quar. Bull. Northwestern U. Med. School, 14:4:258-269, 1940; 15:4:263-269, 1941; 18:1:33-40, 1944. Ann. Otol., Rhinol. and Laryngol., 51:4:891-904, 1942.

Editor's Note: This ms. received in Laryngoscope Office and accepted for publication, July 23, 1946.

in an ossicular chain — converting air waves into liquid waves and these into auditory impulses.

OBSERVATIONS AND DISCUSSION.

In order to appreciate fully the degree to which the ossicle departs from the "standard" pattern of development, it is essential to keep in mind the steps in morphogenesis of a typical long bone. The tibia will serve as an example.

The cartilage "model" of a tibia suggests, in a general way, the form of the future bone: a body, or shaft (diaphysis), ends in slightly enlarged extremities (future epiphyseal areas). Ossification begins in the diaphysis, to produce an elongate collar of bone. Externally, through activity of the osteoblasts in the periosteal covering, the enveloping shell of bone lengthens and thickens; internally, through the action of osteogenic buds which invade the periosteal shell, spicules of endochondral bone and associated marrow tissue come to replace the original cartilage. Bone formation is then augmented by an auxiliary histological mechanism for longitudinal growth, namely, the secondary ossification center. One appears soon after birth. In each, cartilage persists as a growing plate — distally until the individual is 17 years of age, proximally through the nineteenth or twenty-third year. Then, when cartilage is replaced by bone, growth ceases. Cartilage remains only on the articular surface of each extremity. A tibia, then, grows steadily until the individual has reached manhood. In the adult it is 36 times as long as it was when it made a first appearance in cartilage.

In the embryo (*e.g.*, of 28 mm.)* the primordia of the auditory ossicles are branchial in location; the primitive malleus is continuous with the first branchial arch, the incus is near the second; the stapes, seemingly, has acquired independence

*Measurements are of crown-rump length. The ages of this and of succeeding specimens are as follows: 25 mm., 8 weeks; 28 mm., 8½; 50 mm., 11; 111 mm., 15; 117 mm., 16; 126 mm., 16⅝; 135 mm., 17; 146 mm., 17½; 150 mm., 20; 160 mm., 21; 161 mm., 19; 180 mm., 23; 210 mm., 25; 222 mm., 25; 245 mm., 29; 275 mm., 31½; 345 mm., 38.

As will be understood, length does not increase by consistent steps with advancing fetal age, nor are records of age always dependable; evident inconsistencies in relation of length to age (in the above notations) could be due to either or both of these factors.

(see Fig. 1).* They grow only through the first half of intrauterine life, and to only five times the primordial dimensions. When these branchial structures are still cartilaginous, the mandible has begun to form in membrane bone (see Fig. 1). It will ultimately envelop Meckel's cartilage and the latter will be resorbed.

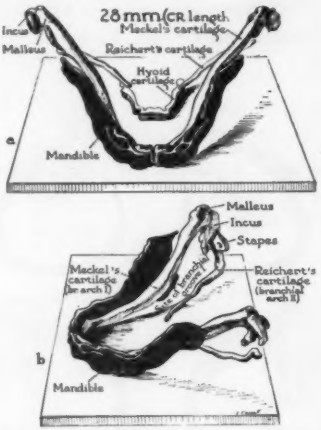


Fig. 1. Reconstruction of cartilaginous ossicles and branchial arches, and the osseous primordia of the mandible in a 28 mm. (CR length) embryo. a. Anterior view; b. superolateral view.

In the fetus of 111 mm. the malleus is broadly continuous with Meckel's cartilage, but the stapes is separate from Reichert's cartilage (see Fig. 3). In the malleus rarefaction of the cartilage is evidenced (see Fig. 3a) — a histological change which precedes the formation of bone. The incus and stapes are as yet unaltered (see Figs. 3b and 3c; levels indicated on inset).

Ossification occurs: first in the incus (117 mm.), next in the malleus (126 mm.), last in the stapes (146 mm.).

*Antecedent stages in the development of the ossicles have been described and figured (in photomicrographs) in earlier papers emanating from this laboratory; the details of origin, which could not be discussed in the present brief account, are being further studied (by means of reconstructions), and will be reported in subsequent articles.

As seen in reconstructions (see Fig. 2), these centers for malleus and incus are established in the 135 mm. fetus (see Figs. 2a and 2b) on the long crus of incus; on the ventral aspect of malleus, at junction with Meckel's cartilage; on the obturator surface of the base of stapes (not included in figure).

Bone has spread from each of these single centers in the 161 mm. stage. The tympanic ring is already an almost complete annulet of bone (see Figs. 2c and 2d).

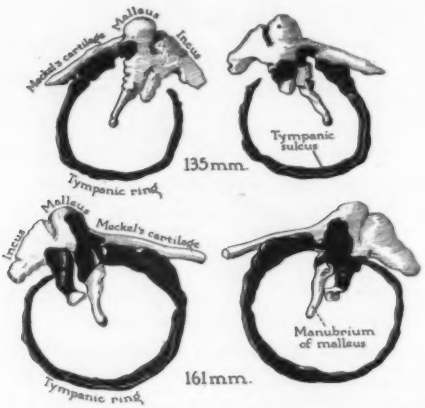
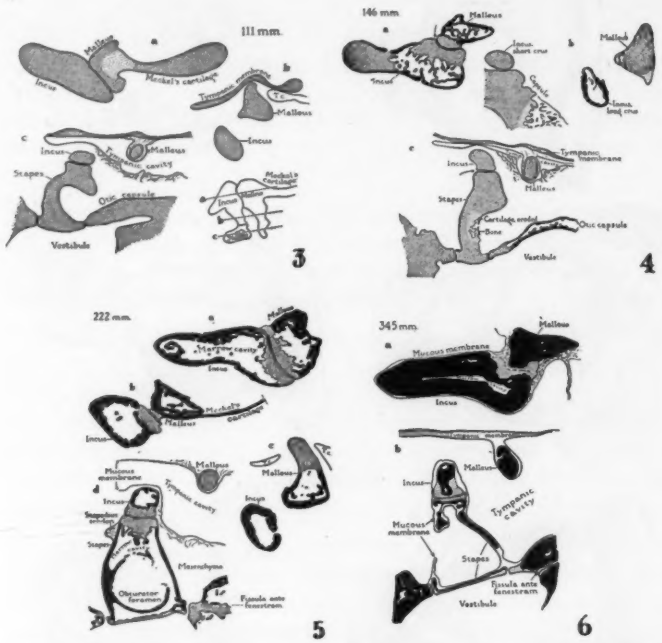


Fig. 2. Reconstruction of malleus (and Meckel's cartilage), incus and tympanic ring. Medial and lateral views. 135 mm. and 161 mm. fetuses.

In the fetus of 146 mm., ossification begins on the obturator aspect of the crus and of the base (see Fig. 4c). At the same time, ossification is well advanced in the long crus of the incus (see Fig. 4b), has not affected the short crus. Perichondral bone covers both ossicles at the level of the articulation (see Fig. 4a). For each ossicle there is a single center of ossification.

At the 222 mm. stage, Meckel's cartilage (see Fig. 5b) is a lancet-shaped bar; it is still continuous with the malleus. In the malleus, endochondral bone is forming. The stapes is not

only deeply excavated (see Fig. 5d), but its obturator wall — capital, crural, basal — has been partially removed by osteoclasts. The original marrow cavity is exposed to the invading mucous membrane. Endochondral bone spread across the cartilage of the base. Later the original perichondral bone will



Figs. 3 to 6. Tracings of sections through the ossicles. Fig. 3, from 111 mm. fetus, at levels indicated in the inset. Fig. 4, 146 mm. fetus at levels similar to those in preceding figure. Fig. 5, 222 mm. fetus. Fig. 6, tracings, 345 mm. fetus. In each figure ordinary stippling represents unaltered cartilage, lighter stippling indicates rarefied (calcifying) cartilage, and black represents bone.

be almost entirely removed; the newly formed endochondral bone is permanent.

By the 345 mm. stage, endochondral bone formation in the malleus and incus has progressed to the point where they

exhibit relative solidity (see Fig. 6a). The stapes, on the contrary, has been reduced to the state of a morphologic

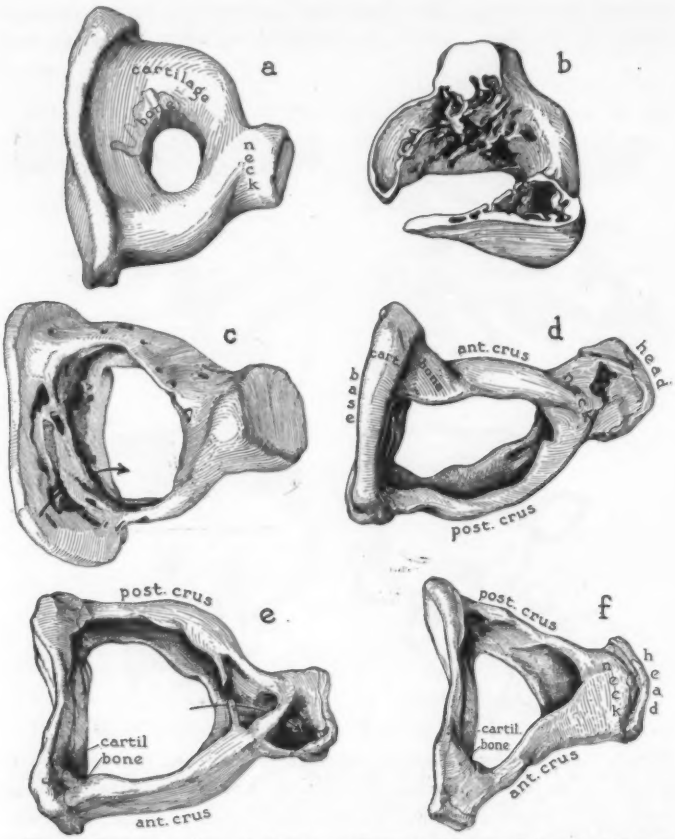


Fig. 7. Reconstruction of stapes. **a.** From a 150 mm. fetus; **b.** 180 mm.; **c,** 210 mm.; **d,** term fetus; **e,** 18-year-old adult; **f,** 57-year. Approximately to scale. In **a** the single ossification is outlined (on the base); in **b** an upper portion has been removed to reveal the extent of hollowing (especially of the crura); in **c** an arrow passes beneath the stapedial crest; in **e** an arrow passes through space in the eroded head.

phantom (see Fig. 6b). It is thin throughout; the obturator, or inner, wall has been sacrificed. Marrow has disappeared.

Mucous membrane now occupies the former site of marrow tissue.

Now to consider, in greater detail, the special features which characterize the development of the stapes.

In the 50 mm. fetus the definitive portions of the stapes are clearly recognizable; at an earlier stage, 25 mm., the stapes is still virtually a ring.

The stapes of the 126 mm. fetus is a cartilaginous element of true stirrup form. A cylindrical head and flattened base join equally robust crura. The intercrural space, or obturator foramen, is relatively small and circular. The tissue is unmodified cartilage.

In the 150 mm. specimen the posterior crus is the more bulky and bowed of the pair, forecasting a regular difference in the adult. The single center of ossification appears on the obturator surface of the base (see Fig. 7a).

Bone spreads rapidly therefrom, toward the head. In the 160 mm. fetus, ossification involves all of the base except the vestibular aspect — which always remains cartilaginous. At the capital end bone has not yet crossed the obturator wall. While bone spreads to produce a complete ring, it is being destroyed on the inner (obturator) surface, left intact on the outer surface.

At the 180 mm. stage, bone has spread to surround the entire obturator foramen. Crura and base are hollowed (see Fig. 7b). Concurrently with continuing ossification, the newly formed bone is removed around the obturator aspect.

The entire obturator wall, in the 210 mm. fetus, has been destroyed by osteoclasis. In the base, some of the perichondral bone persists as a marginal lip, and, in some specimens, as the transverse stapedial crest (see Fig. 7c).

At the 245 mm. stage, adult configuration is almost attained, is actually reached in the 275 mm. fetus. In all specimens cartilage remains on the head and base.

In the term fetus (see Fig. 7d), infant, child or adult (see

Fig. 7e), the head of the stapes may be perforated or uneroded (see Fig. 7f); the margin of the base may be prominently lipped (see Fig. 7d) or flattish (see Fig. 7e), the crura bowed (see Fig. 7e) or relatively straight (see Fig. 7f).*

A stapes, it is now clear, differs strikingly in structure from a long bone. A typical bone increases in size not only through lengthening of its shaft but also through growth at both extremities; its several portions remain externally intact, the shaft merely hollowed to house marrow tissue.

The stapes, on the contrary, beginning as a ring of cartilage, becomes a modified ring of bone — modified to the extent of being flattened in the basal portion, foraminous on its inner aspect and lacking epiphyseal areas. Finally, it resembles a bone that has been split lengthwise, excavated throughout, with cartilage persisting only on the circular head and on the oval base. A tibia continues to grow, into the period of early manhood, while the stapes has attained adult dimensions before the fetus has reached the half-way mark in its intrauterine existence. After the fifth fetal month, changes in the stapes belong in the category of form, not of growth.†

SUMMARY.

1. The ossicles follow a remarkable series of developmental steps in attaining adult form — a circumstance which might be expected when it is considered that they are made over to serve, in higher vertebrates, a function very different from their primitive office.

2. The malleus and incus are developmentally less aberrant than the stapes. Yet they differ from bones generally, not only in possessing peculiar form, but also in lacking epiphyseal centers of growth, in having a single ossification center each, and in attaining adult size and shape in the fetal body.

*These and other stages are figured in the several articles published in the Archives and in that published in the Annals (see footnote ‡, p. 561).

†Consideration of growth and form of the stapes, with fuller discussion of the specialized features of osteogenesis involved, is the subject matter of articles which have already appeared in the Annals and in the Quarterly Bulletin (see footnote ‡, p. 561).

3. The stapes, in addition to displaying the features named above, is unique in the following respects: each crus (corresponding to the shaft of a long bone) becomes channeled to resemble a long bone halved lengthwise, with concurrent sacrifice of its marrow and invasion by mucous membrane and submucosal tissue; the head is likewise excavated, the base reduced to a thin plate of either oval or reniform outline.

4. Since the ossicles attain maturity while the remainder of the skeleton is still fetal, the opportunity for structural alteration is great. Since they are remodeled elements — having been profoundly altered to make up an ossicular chain — variation is to be expected, and does regularly occur.

5. These observations must be taken into account in any appraisal of pathological change in the ossicle. Without knowledge of the kind briefly reported here, normal departure from so-called typical form could be erroneously interpreted as the result of disease.

MANUSCRIPTS INVITED FOR NORTON MEDICAL AWARD.

The book publishing firm of W. W. Norton & Co. announce that they are again inviting manuscripts for submission to be considered for the Norton Medical Award of $3,500 offered to encourage the writing of books on medicine and the medical profession for the layman. The first such award was made to "The Doctor's Job," Dr. Carl Binger's book, published last spring, which gave the Doctor's point of view on his work. Announcement will be made shortly of the winning book for 1946. Closing date for submission of manuscripts this year is Nov. 1, 1946. All particulars relating to requirements and terms may be had by by addressing W. W. Norton & Co., Inc., 70 Fifth Avenue, New York 11, N. Y.

STUDIES ON THE INHERITANCE OF DEAFNESS IN THE PUPILS OF THE CLARKE SCHOOL FOR THE DEAF.*

(a) THE COLLECTION OF FAMILY HISTORIES, PEDIGREES AND AUDIOMETER READINGS.

LOUISE A. HOPKINS, M.S.,†
Northampton, Mass.

(b) THE GENETIC ANALYSIS OF THE PEDIGREE DATA OF THE CLARKE SCHOOL PUPILS.

MADGE T. MACKLIN, M.D.,‡
Columbus, Ohio.

WITH MATHEMATICAL APPENDIX.

PROF. H. B. MANN and RANSOM WHITNEY,
Columbus, Ohio.

No one who is coming in daily contact with deaf children and their parents or with deaf or deafened young people can fail to be extremely concerned and puzzled by the problem of inheritance of deafness. The physician is often at a loss to explain the single occurrence of deafness in a family in which there has been no known history of deafness, or to account for the special predilection for involvement of the middle ear or auditory nerve in certain cases of measles, scarlet fever and other diseases of childhood. What shall the mother or teacher tell the deaf child when complete realization of his deafness first comes upon him? What advice shall be given a former pupil who is contemplating marriage with a former deaf classmate? What can we say to the hearing brothers and sisters when they ask whether they may have deaf children if they marry? These are but a few of the perplexing problems which made it seem wise that one branch of the Research Department at Clarke School be devoted to the

*Read at the Seventy-eighth Annual Meeting of the American Otological Society, Inc., Chicago, Ill., June 1, 1946.
†Research Department, Clarke School for the Deaf.
‡Research Associate, Department of Zoology, Ohio State University.
Editor's Note: This ms. received in Laryngoscope Office and accepted for publication, July 23, 1946.

study of the Inheritance of Deafness in the hope that some of these questions might some day be at least partially answered out of Clarke School's own experience.

Of course you all know that important and far-reaching advances were made in this field by Dr. Alexander Graham Bell, Dr. Edward Allen Fay, Dr. J. Kerr Love and other scientists and educators in this country and abroad, but many of these studies were made before the recent advances in the science of genetics, in otology, in the physics of sound and in a broader understanding of the relation between the ear and pathological conditions in other and sometimes remote parts of the body. These earlier studies were not based on careful audiometric examinations because there were no audiometers and they did not take into consideration the different types of deafness. The problem is still far from solved. This is partly due to the fact that deafness is not a distinct disease entity, but may result from disease, from abnormal or defective development at almost any point along the sound conducting or sound perceiving apparatus, and that any study concerning the inheritance of deafness must in reality be a study of the inheritance of the different types of deafness. It is also due to the fact that recent advances in the science of genetics make us realize that the problem is much more complicated than it may at one time have been thought to be. From some of the charts, you will realize the devious ways in which deafness, even the so-called congenital deafness, appears and disappears in the family tree.

It would be difficult to find a richer field for research along these lines than Clarke School for the Deaf, which was founded in 1867. It is one of the oldest schools in the United States where deaf children are taught to speak and to understand speech. Children enter Clarke School at five years and usually remain over a period of 10 years until they are prepared to enter high school with hearing children. The majority of the children live at the school. We have, from time to time, representatives of old New England families in which there have been accurate records of the occurrence of deafness back over many generations. We have an old and very loyal alumni group, a considerable number of whom, unfor-

tunately, have had deaf children to send back to Clarke School. We have the absolute cooperation of the parents of our children, who are only too eager to do all in their power to help us in our efforts to learn more about the causes of deafness in childhood and the part played by inheritance in the production of deafness. And last, but by no means least, we have the children themselves over a long period of years so that it is possible to study them from many angles, the genetic, the otological and the general medical. In this connection, we are studying the hearing curves not only of the individual child but also of as many members of his family as possible, and we are coming to believe that in many families in which profound deafness has occurred, there are not only deaf individuals and hearing individuals, but also individuals with all gradations of hearing defects, from the almost imperceptible drops in the curves of some to the profound losses in others.

Our first step in the study of the inheritance of deafness has been to collect family histories of present and former pupils of Clarke School. The family history has been taken in the form of a pedigree chart, showing all members and going back at least four generations whenever possible. We have tried to bring out on the charts all possible facts regarding the transmission of deafness, the relation between deafness in childhood and hearing defects in early adult and later life, the relation between deafness and the many other conditions which may or may not have a bearing on the occurrence of deafness. We have noted cancer, tuberculosis, alcoholism, lues, epilepsy, feeble-mindedness, mental disorders, endocrine disturbances, such as goiter and diabetes, and major organic conditions, such as heart trouble or kidney trouble, if these have been predominant in the family history.

The greater part of the data from which the pedigrees were made was obtained through personal interviews with parents and with as many other relatives of our children as possible. These interviews took place in the pupil's home or at the homes of other relatives. Some histories represent a single half-day interview, while others represent numbers of interviews with members from different branches of the family

over a long period of time. Corroborative evidence was obtained from family doctors, social workers and teachers. Other sources of information include reports from hospitals, school records, genealogical records of certain families, information from family Bibles as to births, marriages and deaths as well as the same type of information obtained from church records and records of Massachusetts towns. Finally, some of the data were derived from a community study made in a small, and rather isolated, New England community in which a great deal of deafness had occurred. Here, we were able to interview quite a number of individuals between 80 and 90 years of age who were related to two of our present pupils. Several of these told us many interesting stories of the time when Dr. Bell visited this same community and discussed with them the deafness which was at that time very prevalent there.

The criticism has often been made that people know very little of their forebears, of the cause of death, of the defects they exhibited when alive, of the illnesses from which they suffered and that information obtained from relatives is therefore unreliable. We cannot be sure that each statement obtained from relatives is absolutely accurate, but we do feel that their ability to remember whether or not their relatives were deaf, especially those deaf from childhood, is reliable. Since many of the relatives of our deaf children have been educated in other special schools for the deaf, additional corroborative evidence has been obtained from the records of such schools.

This material added to the original information given by members of the family has yielded data from which very extensive pedigrees could be made. For example, in Pedigree 14, information concerning the individuals in the first six generations and those in part of Generation VII was derived from vital statistics and from published records of family archives. This material was obtained some years after we had derived by means of personal interviews with many relatives of the three deaf pupils in that family, the data used in the rest of Generation VII and the succeeding generations.

Another example is shown in Pedigrees 27 and 124. Some

time after the original information was obtained for Pedigree 27, we found that this pedigree was closely linked with a community study which we were then making. Still later we found that a much younger pupil in Pedigree 124 was related to the one in Pedigree 27 and that both were descended from a common ancestor. The family names were different and the families had never even heard of one another.

When the study was commenced, we were unaware of the part that other defects and diseases might play in contributing to the deafness. For this reason, we noted a history of alcoholism, cancer, insanity, epilepsy and other conditions which appeared in the families. None of this material is used in the abbreviated pedigrees which we will show you, since the data we have accumulated would not suggest that these conditions play a part in the production of deafness; however, if at some future time these conditions give evidence of being related to deafness, the original charts can then be analyzed from that standpoint; thus, it will be seen that we have in our files an enormous amount of data which does not appear on the condensed charts.

The many pedigrees which we have made are not all equally informative, for it has not recently been possible to interview the families of all our pupils in their own homes. When first interviewed at the school, many of the younger parents know very little about their relatives and have never shown much interest in the family history; however, when they realize what we want, they are most cooperative, and the next time they come they will bring with them much more data about the family which they will have obtained from grandparents and other relatives who never visit the school. Information obtained from interviews at the school accumulates much more slowly and is never as extensive or as satisfactory as that obtained from interviews in the home.

Other family histories which are often meager are those obtained from families in which the child's parents were born in a foreign country. Frequently these parents left their homes abroad when young and have heard little or nothing from even near relatives since.

Another group in which the data are scanty is among the

families in which the history is given by parents who are themselves deaf. They are always most cooperative, but their own knowledge is usually extremely limited.

Pedigrees of such families have been made in as much detail as possible, but they are far less accurate and far less inclusive than in the first group. Audiograms have been taken of relatives who visit the school, and when possible reports from hospitals, etc., are obtained later.

The data described above have been used in constructing pedigree charts with legends for each of our pupils. These charts and legends include all the details obtained from all these sources. Each pedigree is given a number in Arabic figures, each generation a number in Roman numerals, and each individual in each generation a number in Arabic figures, so that every individual who ever appears in the files has a distinct and separate number, and the name drops out of sight except during interviews. Below each pedigree chart is a legend giving for each individual, birthplace and date of birth, age at death and cause of death, condition of health and hearing, and general remarks, including occupation and residence, when known.

Each pupil's folder consists of the family history in the form of a pedigree chart, the pupil's own history, including onset of deafness, details of birth, physical development and medical history during preschool years, otologist's report and physician's report of examination after the child enters school, and audiometric readings.

In many cases, these pedigrees are too extensive to reproduce in their original form and for this reason condensed charts have been made for publication. Every effort has been made to include all facts of importance and to exclude unimportant details. It is usual in pedigree charts to list the mates of the various members of the family being studied. This, in a large part, has not been done in these condensed charts unless the mate was deaf. In all cases where the children are shown as descending from only the parent connected with the family in question, the other parent was a hearing person. In the original charts each sibship was arranged in order of birth, with the oldest child on the left and each succeeding

child in order on the right. In the condensed charts that order has frequently been altered so that the males in a sibship are combined in one group and the females in another. This reduces the size of the pedigree to a chart easily readable, and not too long to be handled conveniently. This grouping of individuals was done, however, only where there was nothing pertinent to be brought out. The sibships in which the deaf offspring occur have been completely expanded in each case so that the order of births, miscarriages, etc., is always accurately recorded.

In order to make the direct line of descent easier to follow, the symbols for parents, grandparents and great-grandparents of the deaf child have been outlined more heavily than those for other individuals in the pedigree.

Marriage between related persons is made more readily recognizable by changing the horizontal line, joining a male and a female, indicating a marriage, to a sawtoothed line to indicate a consanguineous marriage. When two related persons are in different generations, the line breaks and descends from one generation to the other. Where a mating line crosses a number of vertical lines which lead to offspring, the line is broken by notches.

We will show you only pedigrees of congenitally deaf children, but there will be several groups shown: some of congenitally deaf children who have hearing parents but other deaf sibs; some of congenitally deaf children who have hearing parents and hearing sibs but in whose family other instances of deafness have appeared in previous generations; some of congenitally deaf children who have deaf parents; some of congenitally deaf children in whose families there are many individuals with slighter hearing defects, and finally, some of congenitally deaf children from families in which there have been consanguineous marriages. These are just a few of the pedigrees which have been compiled since the founding of the Research Department Concerning Heredity of Deafness. Much of the credit for this very fine piece of work should go to the late Dr. Ruth P. Guilder, whose devotion, insight and untiring efforts during her six years in the department have done much to make this work possible.

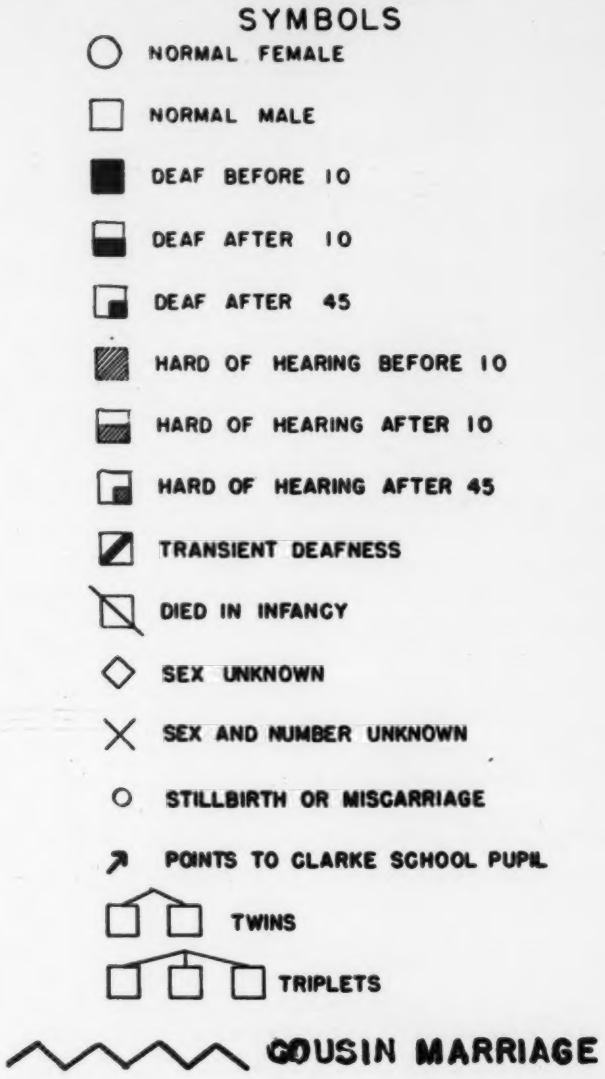
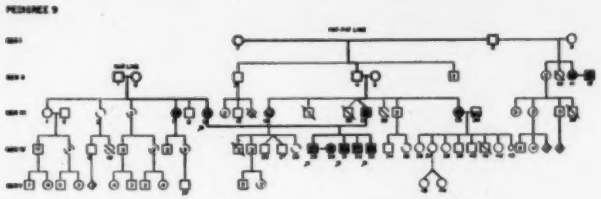
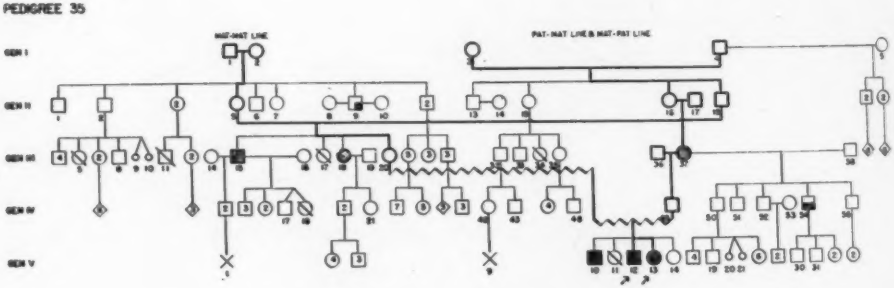

SYMBOLS
NORMAL FEMALE
NORMAL MALE
DEAF BEFORE 10
DEAF AFTER 10
DEAF AFTER 45
HARD OF HEARING BEFORE 10
HARD OF HEARING AFTER 10
HARD OF HEARING AFTER 45
TRANSIENT DEAFNESS
DIED IN INFANCY
SEX UNKNOWN
SEX AND NUMBER UNKNOWN
STILLBIRTH OR MISCARRIAGE
POINTS TO CLARKE SCHOOL PUPIL
TWINS
TRIPLETS
COUSIN MARRIAGE

Fig. 1.



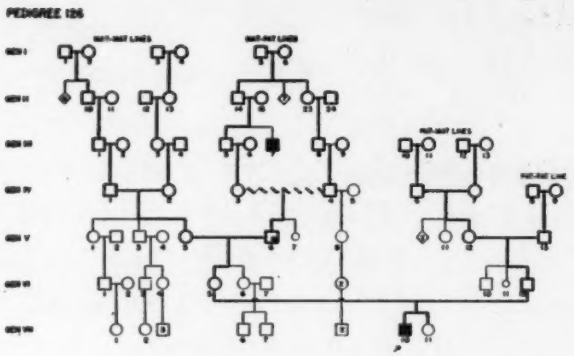
Pedigree 9 — Fig. 2.

II 11 Very hard of hearing since convulsions and fever at three.
12 Born deaf.
III 7 Hard of hearing always.
9 Deafness attributed to scarlet fever at two.
16 Deaf in right ear since measles at five. Low mentality.
21 Deafness attributed to scarlet fever at 18 months. Other twin died.
25 Born partially deaf. Moron.
26 Very hard of hearing since 15 when hit on head by hammer thrower at circus.
IV 29 Born partially deaf.
30 Deaf following illness at two. Possibly meningitis.
31 Born deaf.
32 Hard of hearing since birth.
33 Born deaf.



Pedigree 35 — Fig. 3.

III 15 Hard of hearing after scarlet fever at five.
18 Hard of hearing after scarlet fever at six.
37 Deafness attributed to scarlet fever at one.
IV 54 Hard of hearing from abscessed ears at seven and at thirteen.
V 10 Deafness attributed to cerebral hemorrhage at 18 months. Helpless imbecile.
12 Born deaf.
13 Born deaf.

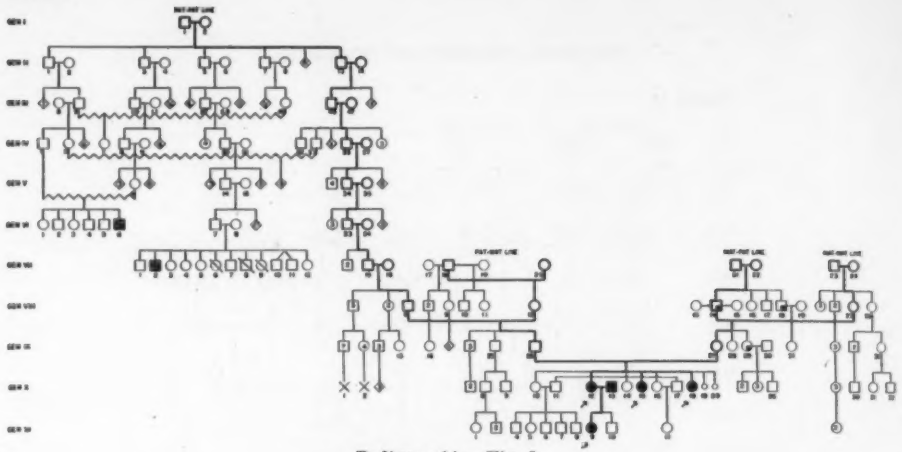


Pedigree 126 — Fig. 4.

III 7 "Deaf mute."
V 6 Slightly hard of hearing since 60's.
VII 10 Born deaf.

Note: It is often said that no one remembers back more than three generations; that data going back further is not reliable. In Figs. 4, 5 and 7 particularly, there were more generations involved than four. In Fig. 5, the information was gathered from volumes of vital statistics of old Massachusetts towns, and did not depend upon hearsay from the family; in Fig. 7, the history of V 1 to V 7 and their descendants was obtained from that family, and the history of V 15 to V 29 and their descendants obtained from that group. The data of the first four generations, and the linking ties between the two family groups were made through vital statistics of these Massachusetts towns. The data in Fig. 4 were gathered from family records.

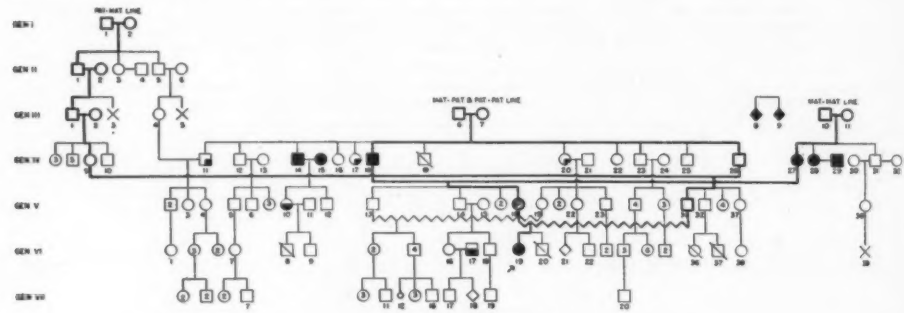
PEDIGREE 14



Pedigree 14 — Fig. 5.

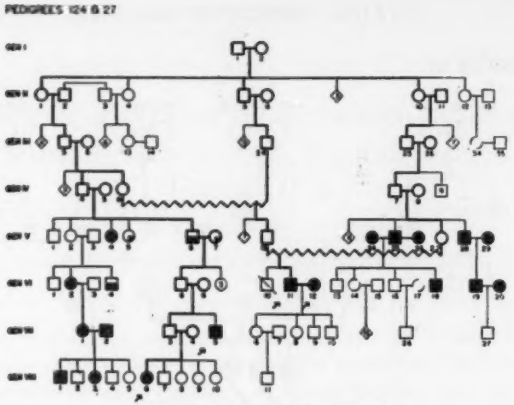
VI	6	Born deaf. Died unmarried at 68.
VII	2	Born deaf. Died unmarried at 23.
VIII	14, 18	Hard of hearing since late 60's. Arteriosclerosis and Bright's disease.
IX	29	Hard of hearing in one ear due to "catarrh." Cerebrospinal meningitis as a baby.
X	12	Born deaf.
	13	Deafness attributed to scarlet fever at three. No deafness in family as far as known.
	15	Born deaf.
	18	Born deaf.
IX	9	Born partially deaf.
	10	Born partially deaf.

PEDIGREE 234



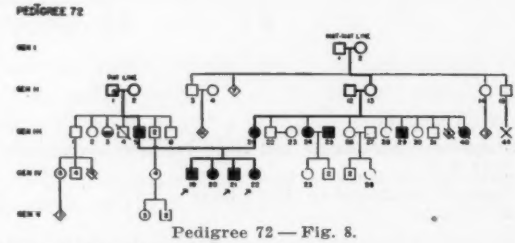
Pedigree 234 — Fig. 6.

III	8, 9	Deaf. No details. Cousins of III 10.
IV	9	Hearing good. First cousin once removed of III 4.
	11	Hard of hearing in later years. Died at 90.
	14	Born deaf.
	15	Born deaf. Had deaf parents and seven deaf brothers and sisters.
	17	Hard of hearing since 50's.
	18	Deafness attributed to whooping cough at five. "Gradually lost his hearing."
	20	Hard of hearing for last 10 years of life.
	27	Born deaf.
	28	Born deaf.
	29	Deafness attributed to scarlet fever at two and one-half.
V	10	Hard of hearing since 20's. Now very deaf.
	18	Always hard of hearing. Married first cousin V 31.
VI	17	Deaf following cerebrospinal meningitis at 17. His aunt became deaf following cerebrospinal meningitis at 10 months.
	19	Born partially deaf. Deafness seemed to increase following scarlet fever at six. Abscesses with scarlet fever. None since.



Pedigrees 124 and 27 — Fig. 7.

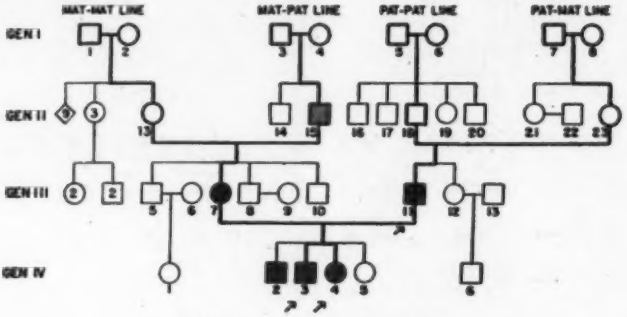
V	4	Deafness attributed to scarlet fever at 18 months. Died of tuberculous abscess at 70.
	5	Slightly hard of hearing in later years. Died at 66.
	6	Hard of hearing since Civil War. Now 93.
	24	Born deaf. Died of apoplexy at 36.
	25	Born deaf.
	26	Deaf. Cause unknown.
	28	Born deaf.
VI	2	Hard of hearing since scarlet fever at six. Now, at 70, very deaf.
	4	Deafness attributed to injury of ear at eight. Has been "totally deaf" for past 20 years.
	11	Born deaf or deaf following scarlet fever at eight months.
	12	Deaf following meningitis at three. "Sang and talked before she had meningitis."
	18	Born deaf. Died of diphtheria at 11.
	19	Deafness attributed to scarlet fever at seven.
	20	Born deaf.
VII	1	Born deaf.
	2	Deafness attributed to "brain fever" at 20.
	5	Born deaf. Insane since 23.
VIII	1	Born deaf.
	3	Born deaf.
	6	Born deaf.



Pedigree 72 — Fig. 8.

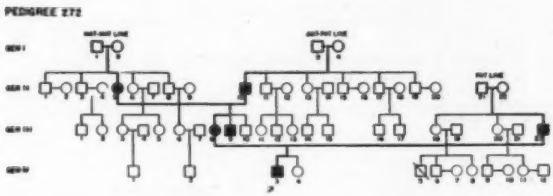
II	1	Hard of hearing after 60. Died at 80.
III	3	Hard of hearing since 30's.
	5	Born deaf.
	21	Born deaf.
	24	Born deaf.
	25	Deaf. Cause unknown.
	29	Born deaf.
	40	Born deaf. Died of convulsions at one.
IV	19	Born deaf.
	20	Born deaf.
	21	Born deaf.
	22	Born deaf.

PEDIGREE 21



Pedigree 21 — Fig. 9.

II 15 Hard of hearing when young. Killed at 46.
III 7 Born deaf.
11 "Became totally deaf following eruption of teeth at 20 months." Nervous and unstable.
IV 2 Born deaf. Died of septic poisoning at 14.
3 Born deaf.
4 Born deaf.



Pedigree 272 — Fig. 10.

II 5 Deafness attributed to "brain fever" at two.
10 Deafness attributed to "fall from wagon at seven."
III 8 Born deaf.
9 Deafness attributed to "sickness in infancy."
22 Deafness attributed to "brain fever at two."
IV 3 Born deaf.

(b) GENETIC ANALYSIS OF DATA AND PEDIGREES.

MADGE T. MACKLIN, M.D. (by invitation),†
Columbus, Ohio.

WITH MATHEMATICAL APPENDIX.

PROF. H. B. MANN and RANSOM WHITNEY,
Columbus, Ohio.

Briefly, I will discuss four questions arising from an analysis of these data. There is among the pupils at Clarke School a large group of deaf children in whose family histories there is no other case of deafness; these are the only persons known to be deaf in the entire family. Some of these children are among those known to be deaf because of some extrinsic cause, such as infection or injury. Some are deaf because of the occurrence of German measles in the mother during the early months of the pregnancy resulting in the deaf child; some children have been recognized by the parents as having been born deaf, since they discovered the deafness early in infancy before any illness had occurred which could account for it. There remains a large group of isolated or so-called sporadic cases of deafness, attributed by the parents to some illness which the child suffered in early infancy before the fact was established that the child heard because it used intelligible words or phrases. The first question to be answered is this:

1. What proportion of this group of isolated cases of deafness can we safely assume were hereditary cases? We know that there must be families in which only one child is deaf, and in human families which are small, this group will form the majority. What proportion can we assume are really deaf through some extrinsic cause?

2. What mode of inheritance do we find among the pupils

*Read at the Seventy-eighth Annual Meeting of the American Otological Society, Inc., Chicago, Ill., June 1, 1946.
†Research Associate, Department of Zoology, Ohio State University.

of the Clarke School for the Deaf, when deafness is probably congenital, and of the nerve deafness type?

3. Is there any evidence from these families that there is more than one type of congenital nerve deafness?

4. Is there any evidence that a child may be genotypically deaf, but phenotypically hearing?

Question 1: Before this question can be answered we must eliminate from our group of deaf pupils the following cases: *a.* Those in whom there is clear evidence that the child heard before the onset of the illness which was said to be the cause of the deafness. Inasmuch as most parents do not test the hearing of their children, and may mistake facial responses on the part of the child as evidence of hearing, when in reality they may merely be responses to familiar faces and objects, what criterion can we use to be certain that the child was born hearing? Three groups of children were classified as deaf through infection. Here the excellent medical histories were of inestimable value. If the child has spoken in intelligible words or phrases, then suffers from an illness after which its speech faculties are lost, we may safely assume that the illness caused the deafness. Second, if the child had spinal meningitis, as revealed by opisthotonos, bacteriologic confirmation, lumbar puncture, etc., it may be reasonably assumed that the deafness was caused by the disease, even though the illness occurred before the child had been speaking. Third, a bilateral operation for mastoiditis was accepted as a cause of deafness.

b. The second class of sporadic cases to be eliminated are those in which a history is obtained of German measles in the mother during the early months of the pregnancy which ended in the birth of the deaf child.

c. All families were eliminated in which one or both parents were deaf. This was done for two obvious reasons; the first being that when the parents are deaf the deafness in the child does not have to be proved to be hereditary, it can safely be assumed to be so in the vast majority of cases. The second is that the formula used in the analysis is dependent upon

the assumption that the parents of the deaf child are both hearing. After these groups were omitted, all other cases of deafness in which the parents were hearing were grouped together; the assumption was made that all the cases of deafness were hereditary, and the data analyzed on the theory that the deafness was due to a recessive gene substitution.

TABLE 1. FAMILIES WITH DEAFNESS PRESUMABLY HEREDITARY IN THE CHILDREN.

Number of Sibships	No. of Sibships with Only One Deaf Child	No. of Sibships with Only One Deaf, No Other Case of Deafness in Pedigree	No. in Which Child Was Known to Be Born Deaf	No. in Which Deafness Was Attributed to Illness
272	214	172	78	94

The data are tabulated in Table 1. There were 272 sibships, and of these, 214 had only one deaf child. In 42 of these, the deafness was recognized by the parents or the author as probably hereditary, inasmuch as there were other cases of deafness in other sibships in the family. In the remaining 172 sibships, there was no other case of deafness in the family pedigree, so that the deafness might possibly be considered to have been caused by infection. In these 172 sibships, the parents recognized that the child was born deaf in 78 instances, and made no attempt to attribute the deafness to some extraneous cause. That left 94 sibships in which the deafness was not considered to have been present at birth, and to have been caused by some illness which the child had suffered in early infancy which prevented its ever learning to speak. The question with which we are now concerned is this: what proportion, if any, of these 94 deaf children should we consider as really belonging to the hereditarily deaf, and what proportion are probably deaf because of some infection suffered by them or by their mothers during the pregnancy in which they were carried?

There appears to be no adequate statistical method by which this percentage can be derived. We can examine matings of deaf with deaf, which produce all deaf children, and in which we are justified in assuming that the deafness was hereditary. There were nine such matings in these pedigrees,

with some statement made as to the cause of deafness in one or both parents. Of these 18 deaf parents whose progeny showed them to be hereditarily deaf, 12 were said to have been deaf through infection. This means that in this small group, two-thirds of the persons have erroneously been claimed to have been deaf through infection, when in reality they were hereditarily deaf. We cannot state that this percentage would be found in all groups of doubtful cases of infectious deafness or that it applies to the group of 94 families which we have here.

The objection may be raised that I have no proof that 66 per cent or any per cent of these children whose deafness appeared to be an isolated case were in reality inherited, but I should like to state the following facts in support of the suggestion that some percentage of these cases should be transferred from the infectious to the hereditary group.

1. Many parents state that their first deaf child's deafness was caused by measles, etc., sometimes that a second deaf child's deafness was caused by whooping cough, etc., but when a third child is deaf they will be forced to admit that the deafness in that child was not caused by any illness. The logical assumption is that the deafness of the other children in the family was not caused by illness either (see Fig. 2, III 16, 21, 25).

2. Many of these people whose deafness their parents have said was caused by infection marry other deaf people whose deafness was also attributed to infection, and produce all deaf offspring, thus indicating the hereditary nature of their deafness (see Fig. 2, III 9, III 21).

3. Measles, whooping cough, etc., seldom cause deafness in children who are old enough to have proved their ability to hear by speaking in intelligible words and phrases. From this we must conclude either that *a.* the cochlear cells are much more susceptible in the infant or young child than in one who has learned to speak; or *b.* that these diseases do not cause more deafness in the infant than in the child who has learned to talk.

4. Many children with proved infectious diseases in early infancy have undamaged hearing.

5. The analysis of families (see Table 4) in which deafness

TABLE 2. ANALYSIS OF CONGENITAL NERVE DEAFNESS ON THE ASSUMPTION THAT IT IS CAUSED BY A RECESSIVE GENE.

No. in Family	No. of Families	Tot. No. of Children	Tot. No. Deaf	Tot. No. Expected to Be Deaf	Standard Deviation Squared
1 (1)	37	37	37	37	0.00000
2 (1)	46	108	62	61.7112	6.61230
(2)	8				
3 (1)	51	171	64	73.9461	14.98929
(2)	5				
(3)	1				
4 (1)	26	160	54	58.5120	16.80200
(2)	14				
5 (1)	22	160	45	52.4448	18.93696
(2)	7				
(3)	3				
6 (1)	8	78	21	23.7224	10.08735
(2)	2				
(3)	3				
7 (1)	11	119	26	34.3332	16.49408
(2)	4				
(3)	1				
(4)	1				
8 (1)	6	64	11	17.7800	9.37920
(2)	1				
(3)	1				
9 (1)	4	72	14	19.4624	11.04160
(2)	3				
(4)	1				
11 (1)	1	22	3	5.7420	3.61160
(2)	1				
12 (1)	2	48	8	12.3920	8.07840
(2)	1				
(4)	1				
Total	272	1039	345	397.0461	116.03278
*Total	235	1002	308	360.0461	116.03278

*Elimination of all one-child families, where observed and expected values must agree, gives the second set of Total Values.

$\sigma^2 = 116.03278 \therefore \sigma = 10.8$

$360 - 308 = 52$, which is more than 2σ or 21.6

All children dying in infancy, also all miscarriages and stillbirths, were omitted from these families. The numbers in parentheses in Column 1 indicate the number of children deaf in the family.

Note that in all the sibships except those with one and two children, the observed number of deaf is always less than the expected number. Agreement between these two values is, of course, always perfect in the one-child families, as the expectation is always 100 per cent.

could be recognized as running through the family was in close agreement with the theory of recessive inheritance. There is no reason to assume that those families in which several children were deaf, but in which no other deafness was known, had any different type of deafness from these families. One, therefore, requires a group of families with only one deaf child, some of whom must come from the 94 families with a child supposedly deaf through illness.

Question 2: What mode of inheritance is exemplified in this group of families with congenital nerve deafness at Clarke School?

We shall test the hypothesis that the deafness in these families in which the parents are hearing is dependent upon a recessive gene. If this is true, then we should expect that the chances of a child's being deaf in such matings are one in four, while the chances of a hearing child are three in four. The group of families must be separated into the sibships with two, three, four, etc., children as in Table 2. The chances of having families with one and with two children affected may be expressed by the binomial theorem, with **la** or just **a** representing the chance of being deaf, and 3n the chances of being normal. This expression, expanded to the second, third, etc., powers gives the number of children who will be affected in each size sibship. Thus $(a + 3n)^2 = a^2 + 6an + 9n^2$. This means that for every two-child family in which there are two children deaf, (a^2), there will be six families with one deaf and one normal (6an) and nine families in which both children hear $(9n^2)$. The latter group of families will not be entered in this series, since they do not have a child in the School for the Deaf; the only families who will have pupils in the School will be those with one or both children deaf. There are seven such families, with a total of eight deaf children; two being in the family with both deaf, the other six being in the six families with one only deaf. Since seven families have eight deaf children, the average number of deaf expected on this hypothesis in two-child families is 1.14 or 8/7 deaf children per family. There were 54 such families, so that 61.7112 deaf children would be expected on the basis

of the theory. There were actually 62 deaf children in these families.

As the size of the family increases, the number of deaf children per family increases, since we are only considering those families in which at least one deaf child occurs; we omit all the families in which there were no deaf children. In three-child families, the expectation is as follows: $(a + 3n)^3$ gives $a^3 + 9a^2n + 27an^2 + 27n^3$. Again the 27 families with no deaf children are omitted from the calculations, and we have a total of 37 families with three children: of these, one (a^3) has all three deaf; 9 ($9a^2n$) have two deaf and one hearing; and 27 ($27an^2$) have one deaf and two hearing. The total number of deaf in these 37 families is 48, with an average of 1.2973 per family. This multiplied by the 57 families found at Clarke School with three children, some of whom were deaf, gives the number of deaf expected in these families. The expected value was 73.9461, whereas actually, there were only 64 deaf in this group. Similar computations are carried throughout the table. The total values are 1039 children in 272 families in the records, of whom 345 were deaf, whereas 397.0461 were actually expected to be deaf in this group. This means that we found too few deaf in this group to be in accord with the expectation. There are several explanations for this.

1. We may have included a group of families in which there was but one child deaf, and in which the deafness was not hereditary but caused by some other factor. This inclusion of families with but one deaf, where we would expect more than one to be affected has lowered the incidence of the deafness in this group beyond the expected level.

For example, the formula just given for three-child families was $a^3 + 9a^2n + 27an^2 + 27n^3$. There should be on the average three times as many families with one deaf as with two deaf in this size sibship, and nine times as many with two deaf as with three. Reference to Table 2 shows that there were five families with two deaf, and 51 with only one deaf, whereas one would expect only 15. Substituting the value of 15 for 51 shows that out of a total of 21 families

(15 + 5 + 1) there would have been 28 (15 + 10 + 3) deaf, and that 27.2433 (21 × 1.2973) would have been expected to be deaf on the theory that deafness is due to a recessive gene. The inclusion in the group of 36 families with one child deaf has lowered the observed value well below the expected.

2. The deafness we are dealing with here may have modes of inheritance in some families, at least, which call for fewer deaf than one in four when the parents are both hearing. Such a mode of inheritance would be one in which the deafness was dependent upon the simultaneous presence of two recessive gene pairs in the chromosomes, so that with both parents heterozygous for both genes, the expectation of deaf would be not one in four, but one in 16.

The latter assumption can be tested in the following manner. If the latter theory of two recessive genes is correct, we should find that although we eliminated all families with only one deaf child we would still have too few deaf in our families, if we tested them on the idea that a one in four ratio was correct. This has been done in Table 3, and here after the omission of all families in which only one deaf child occurred, we find that in 50 families, with a total of 283 children, there were 115 deaf and 121.77 who were expected to be deaf. This is an extremely close agreement, the difference between the two values being only 1.4 times the standard deviation. This table suggests that the theory of a single recessive gene as the cause of deafness in any one family is correct, and that the lack of agreement in the entire series of families in Table 1 was not caused by dependence upon more than a single recessive gene.

A third reason for there being too few deaf observed to agree with the theoretical expectation is the small number of families with three or more deaf children.

I have shown that the theory of a recessive gene being responsible for the type or types of congenital deafness here observed, although not *proved* to be true, is in accord with the facts set forth in Table 3.

If we tabulate all families in which the congenital deafness was almost certainly hereditary because deafness existed in more than one sibship, we should have a fair agreement between observed and expected values, if the theory of a recessive gene causing the deafness is correct. This has been done in Table 4. There are 62 families, with a total of 293 children. Of these, 88 were deaf and 95.7 were expected to be deaf. The standard deviation was 6.1, so that the observed and expected values differed by about 1.2 times the standard deviation. This is a good agreement, and is in accord with the theory of recessive inheritance.

TABLE 3. NUMBER OF DEAF CHILDREN IN SIBSHIPS BORN OF HEARING PARENTS.*

No. in Family	No. of Families	Total Children	Expected Value	Observed Value	σ^2
3 (2)	5	18	12.60	13	.54
(3)	1				
4 (2)	14	56	30.94	28	2.66
5 (2)	7	50	23.30	23	3.10
(3)	3				
6 (2)	2	30	12.30	13	2.25
(3)	3				
7 (2)	4				
(3)	1	42	15.54	15	3.60
(4)	1				
8 (2)	1	16	5.48	5	1.56
(3)	1				
9 (2)	3	36	11.56	10	3.80
(4)	1				
11 (2)	1	11	3.23	2	2.70
12 (2)	1	24	6.82	6	3.12
(4)	1				
Total	50	283	121.77	115	23.33

*This table is the same as Table 2 except that all families with only one child affected are omitted. For explanation see text.

$\sigma = \sqrt{23.33} = 4.8$

$121.77 - 115 = 6.77$, which is $1.4 \times \sigma$

If deafness of the congenital nerve variety is hereditary and dependent upon a recessive gene we would expect cousin marriages to be more common among the parents of these deaf children than among the population in general. This we find to be true, the incidence of cousin marriages being about 8 per cent in these families, whereas it is only about 0.2 per cent in the population at large (see Figs. 3, 4, 5, 6, 7).

When cousins marry, they probably have the same type of deafness, so that one would expect a normal ratio of deaf children to appear in proportion to the size of the sibship. These matings are shown in Table 5, and it is clear that there is almost perfect agreement between the theoretical expectation of deaf and the actual number of deaf children observed.

TABLE 4.

No. in Family		No. of Families	Total Children	Expected Value	Observed Value	σ^2
1		5	5	5.0000	5	0.00000
2	(1)	9	20	11.4280	11	1.22450
	(2)	1				
3	(1)	6	27	11.6757	13	2.36673
	(2)	2				
	(3)	1				
4	(1)	5	28	10.2396	9	2.94035
	(2)	2				
5	(1)	7	60	19.6668	19	7.10136
	(2)	3				
	(3)	2				
6	(1)	4	30	9.1240	7	3.87975
	(3)	1				
7	(1)	1	42	12.1176	14	5.82144
	(2)	3				
	(3)	1				
	(4)	1				
8	(1)	1	16	4.4450	3	2.34480
	(2)	1				
9	(2)	1	18	4.8656	6	2.76040
	(4)	1				
11	(2)	1	11	2.8710	2	1.80530
12	(1)	2	36	9.2940	4	6.05880
	(2)	1				
Total		62	293	95.7273	88	36.30343

This table is based upon all families in which there is deafness in more than one sibship, thus indicating the hereditary nature of the deafness.

$\sigma^2 = 36.30343$ $\sigma = 6.1$

95.7 — 88 = 7.7, which is only 1.2 times the standard deviation, 6.1

Although the agreement between expected and observed values of deaf is very close in Tables 3, 4 and 5, as pointed out in the mathematical appendix prepared by Mr. Ransom Whitney under Prof. H. B. Mann's directions, other ratios such as 3/16 or 5/16 in place of 1/4 might also be possible with the values here found. On the other hand, no ratio of

5/16 or of 3/16 is possible unless more than one gene is involved in producing deafness in any one case, and even then the ratios of 5/16 and 3/16 would not be stable; but would vary from 1/4 to 1/2 in the first instance and from 1/16 to 4/16 or 1/4 in the second, depending upon the frequency the two genes in the population.

TABLE 5. NUMBER OF DEAF CHILDREN IN SIBSHIPS IN WHICH THE PARENTS WERE RELATED.*

No. in Family	No. of Families	Total No. of Children	No. Expected Affected	No. Affected	σ^2
1	3	3	3.0	3	0.0000
2	4	8	4.56	5	.4898
4	4	16	5.84	7	1.6802
6	2	12	3.66	3	1.5519
7	1	7	2.02	2	.9702
8	2	16	4.44	2	2.3448
Total	16	62	23.52	22	7.0369

*Those dying in infancy and all miscarriages and stillbirths were omitted. Only sibships in which both parents were hearing were used. The number of families is small, but the number of deaf expected in all cases agrees closely with the number actually observed. This would indicate that no matter what types of deafness were exhibited in these particular families, they were probably all dependent upon recessive genes.

$\sigma^2 = 7.0369$

$\sigma = \sqrt{7.0369} = 2.65$

$23.52 - 22 = 1.52$, which is only 57 per cent of the standard deviation.

The conclusion reached for the mode of inheritance of deafness in these families is that the data here presented in families in which the parents are related or in families in which at least two children are affected, or for families in which more than one sibship is affected, are in accord with the theory of dependence upon one recessive gene. Other possibilities cannot be excluded, but they are genetically not so likely, since they are not such stable ratios.

On the basis of a recessive mode of inheritance, combinations of parents and children are possible other than the ones just analyzed, namely, two hearing parents with some deaf children. These are *1.* two deaf parents; *2.* one deaf and one hearing parent not a carrier, and *3.* one deaf and one hearing parent who is a carrier. Combinations of infectiously deaf parents are also possible. If both parents are deaf with the same type of hereditary deafness, one expects all their children deaf. If one parent is hereditarily deaf and one parent a

normal hearing person, none of the children will be deaf. If the hearing parent is a carrier of deafness the expectation is one-half the children hearing, one-half deaf. Remembering that a person who is deaf due to infection might also be a carrier of hereditary deafness or be genetically a hearing person, we can see further that *4.* two deaf persons, both deaf from infection, might have all hearing children, if neither was a carrier, or *5.* some hearing, some deaf children if both were carriers. Finally, *6.* an hereditarily deaf person mated with a carrier, deafened by an infection, could have some deaf, some hearing children.

In this group of Clarke School pedigrees collected between 1930 and 1940, there were 40 matings of deaf with deaf persons. Six of these matings were childless at the time of the collection of the data; 15 had all deaf offspring (possibility 1) (see Fig. 5, X 12, 13; Fig. 8, III 5, 21; Fig. 2, III 9, 21); 11 matings resulted in all hearing children (possibilities 4, 5, 6) (see Fig. 8, III 24, 25; Fig. 7, VI 11, 12 and VI 19, 20). And finally eight matings had some deaf and some hearing (possibilities 5, 6) (see Fig. 7, VII 1, 2; V 24, 25; Fig. 9, III 7, 11; Fig. 10, III 8, 22).

Question 3: Are there more types of hereditary nerve deafness than just one? If recessive deafness is to appear in a child, it is necessary that a person hereditarily deaf mate either with a person suffering from a similar type of deafness, or with a carrier of the same type of deafness. Should both parents be hereditarily deaf, but deaf with different types of deafness, the children will be hearing, since the types of deafness will not match. One might conceive of two types of nerve deafness, one interfering with the nerve cells of the cochlea, the other interfering with the nerve fibres, yet each dependent upon its own specific gene, and each gene being a recessive one. Two such persons mating would produce all hearing children, each carrying a gene for each type of deafness, yet themselves hearing. Not unless persons with different types of deafness chanced to marry and reveal such a condition would one be aware that the types of deafness in the population differed. Thus, Lindenov in Denmark feels that

there is but one type of nerve deafness, as he has found no evidence to the contrary. It should be pointed out that if the different types were all recessive, and if the various types never happened to mate with each other, there might be a variety of congenital nerve deafnesses, each dependent upon a different recessive gene, and one would never know of the different varieties. The only way they would come to light would be as in Pedigree 234 (see Fig. 6), where two persons (IV 18 and 27), each apparently hereditarily deaf because they had deaf sibs, married and produced a child who was not congenitally deaf, although it was hard of hearing. This child mated with a first cousin (V 31), who it must be presumed had a gene similar to one of the two kinds carried by the first person, and these two hearing people then produced a deaf child (VI 19). The picture may be presented as follows. One deaf person was deaf because of the presence of gene dd; and the second was deaf because of the presence of gene d′d′. Their child was hearing because it was DdD′d′. This person then mated with a cousin who was Dd, the two genes d, d met in the same child and produced deafness. This family shown in Fig. 6 suggests strongly the presence of at least two types of congenital nerve deafness. They cannot be distinguished by audiograms, but can on the basis that although hereditarily deaf, these parents produce hearing children.

Question 4: Can a child be genotypically deaf, but phenotypically hearing? Not until we have more evidence of what happens when hearing children of two deaf parents who have also produced some deaf children marry, and in sufficient combinations of hearing with deaf and with other hearing to afford us adequate data will we be able to attempt the answer to this question.

Theoretically such a thing as being genotypically deaf but phenotypically hearing is possible. Hearing persons might actually have both matching genes for deafness, and in reproducing, would act in the same manner as a deaf person; but through the presence of modifying factors, the level of residual hearing which is always present in these children, although

they are deaf to the spoken voice, might be raised until they could hear sounds with the unaided ear. For example, in diabetes, although the person has the gene for diabetes, he does not show the disease until the critical threshold is crossed, at which 90 per cent of the islet tissue in the pancreas is destroyed. He is a potential diabetic but not exhibiting the disease. So the deaf person might be genetically deaf but still have enough hearing to pass for a hearing person.

This explanation is, of course, not the only one as pointed out above when two deaf persons have some deaf and some hearing children. The hearing children may be considered as illegitimate; one of the parents may be regarded as hereditarily deaf, the other as a carrier but deafened by infection, or still another possibility is that one of the deaf parents who might have been the only deaf person in his family was deaf due to a dominant mutation, and that his deafness is transmitted to about half his offspring, the other half of whom hear. The families shown in Figs. 9 and 10 are ones which might possibly have this latter explanation, or the ones which postulate a person whose residual hearing is enough to raise him into the hearing class, although he is genotypically deaf.

Summary: Experience teaches us that not all cases of deafness which are attributed by parent or physician to some infectious disease in childhood are necessarily so to be explained. On the basis of analysis of families of pupils at the Clarke School for the Deaf, as many as 66 per cent of such cases may be actually examples of isolated instances of hereditary deafness in that family. This should make us cautious in assuring deaf persons whose deafness is presumably caused by infection that their children will not be deaf.

Although the data do not exclude other ratios of deaf to hearing children, they are in agreement with the theory that congenital nerve deafness in these families is dependent upon the presence of a recessive gene.

There is evidence that there may be more than one type of congenital nerve deafness, distinguishable not by audiograms, but on the basis of genetic behavior. These types can be dis-

tinguished only when two obviously hereditarily deaf persons mate, and produce all hearing children.

There is a suggestion in some of the pedigrees of these families that a person may have inherited both genes for nerve deafness, but have enough residual hearing to be classified as a hearing person.

At the invitation of Dr. Frank Reiter, Principal of the Clarke School for the Deaf, Dr. M. T. Macklin, then Assistant Professor in the Department of Histology and Embryology under Prof. Chas. C. Macklin at the University of Western Ontario Medical School, London, Ontario, spent parts of several vacations at the Clarke School going over the data which had been collected from 1930 to 1936 by Miss Louise Hopkins and the late Dr. Ruth Guilder, and from 1936 to 1940 by Miss Hopkins alone. It is this group of data which is briefly summarized and evaluated in this paper. I express my thanks to the Clarke School Trustees and to Dr. Frank Reiter for their permission to report part of the work at this time. I also express my warm gratitude to Prof. Charles C. Macklin, whose unfailing interest in my work enabled me to undertake this research when I was on his staff.

I am indebted to Prof. H. B. Mann, Ohio State University, Department of Mathematics, for his generous advice, and to Mr. Ransom Whitney, who prepared the mathematical appendix to this paper.

MATHEMATICAL APPENDIX.

PROF. H. B. MANN and RANSOM WHITNEY,

Dept. Mathematics, Ohio State University.

1. In the foregoing study of inheritance we are concerned with families where there are at least two children and at least one child is affected. The data from Table 2 are summarized here.

$$\begin{array}{llll} N_2 = 54 & N_6 = 32 & N_8 = 8 & N_{11} = 2 \\ N_3 = 57 & N_6 = 13 & N_9 = 8 & N_{12} = 4 \\ N_4 = 40 & N_7 = 17 & N_{10} = 0 & \\ M = 308 & & & \end{array}$$

where, for example, N_6 stands for the number of families with six children, and M stands for the total number of affected children in all the families.

We wish to test the hypothesis that the probability of a child's being affected in the universe of all children from heterozygous parents is 1/4. This will be referred to as the null hypothesis. We would also like some criterion as to just how good our test is.

2. The Test. If we denote by M the expected value of the total number of affected children under the null hypothesis we have the formula:

$$1.\quad \overline{M} = 1.14N_2 + 1.30N_3 + 1.46N_4 + 1.64N_5 + 1.83N_6 + 2.02N_7 + 2.22N_8 + 2.43N_9 + 2.65N_{10} + 2.87N_{11} + 3.10N_{12}$$

and if σ is the standard deviation of the total number of affected children we have:

$$2.\quad \sigma^2 = .12N_2 + .26N_3 + .42N_4 + .59N_5 + .78N_6 + .97N_7 + 1.17N_8 + 1.38N_9 + 1.59N_{10} + 1.81N_{11} + 2.02N_{12}$$

Now using the values of N_3, N_5, N_4,—from the experiment we find that $\overline{M} = 360$ and $\sigma = 10.7$.

The sample is large enough to approximate the distribution of M by a normal distribution; therefore, 95 per cent of the sample values of M will lie in the range $360 \pm 2\sigma$, that is, from 339 to 381. Any value of M outside of this interval lies in the region of rejection of our hypothesis. The region of rejection of the null hypothesis is called the critical region.

Our test for acceptance or rejection of the null hypothesis will then be: we reject the null hypothesis if our sample falls in the critical region and we accept it if it is not in the critical region.

The experimental value of M was 308. Since 308 is in the critical region we reject our hypothesis of probability 1/4.

This may be due to the inclusion of children whose deafness is due to infection or other accidental causes.

3. In an effort to make certain that in the families that have been selected the parents actually carried the gene for deafness, we will use only those families with three or more children where there are at least two children affected. The principal difficulty will come from our decreased sample size.

The new data summarized from Table 3 are given here.

$N_3 = 6$	$N_6 = 5$	$N_9 = 4$	$N_{12} = 2$
$N_4 = 14$	$N_7 = 6$	$N_{10} = 0$	
$N_5 = 10$	$N_8 = 2$	$N_{11} = 1$	
$M = 115$			

This time the expected value of the total number of affected children and the standard deviation are given by

$$3.\ \overline{M} = 2.10N_3 + 2.21N_4 + 2.33N_5 + 2.46N_6 + 2.59N_7 + 2.74N_8 + 2.89N_9 + 3.06N_{10} + 3.23N_{11} + 3.41N_{12}$$

$$4.\ \sigma^2 = .09N_3 + .19N_4 + 31N_5 + .45N_6 + .60N_7 + .77N_8 + .95N_9 + 1.14N_{10} + 1.35N_{11} + 1.56N_{12}$$

Substituting the values for the N's we get

$$\overline{M} = 122 \qquad \sigma = 4.7$$

Our critical region will again be the value of M outside the interval $122 \pm 2\sigma$, that is, from 113 to 131. This time our sample value of 115 does not lie in the critical region, and we do not reject our hypothesis of probability 1/4.

4. Power of the Test. Now we ask the following question. Although our experiment told us not to reject our hypothesis 1/4, can we be safe in accepting it? Suppose some other probability like 3/16 is actually correct. What is the probability that our sample value of M will fall in the critical region? We certainly want it to do so if 3/16 is correct and 1/4 wrong If this probability is low we say the power of the test with respect to the alternative hypothesis 3/16 is low. Roughly it means that our test offers little chance of distinguishing between the hypothesis 1/4 and 3/16. On the other hand, if the power of the test is high with respect to an alternative hypothesis we can accept our hypothesis with more confidence since we would know that it had a favorable chance for rejection if it were incorrect.

We will illustrate with a graph for our test.

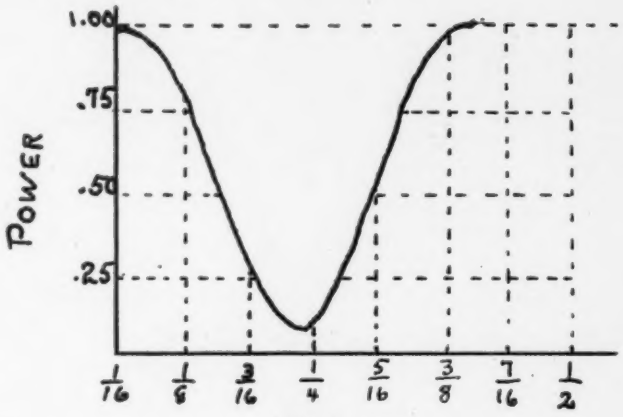


Fig. 1.

Example: For the alternative hypothesis 5/16 it was found by calculation that if 5/16 were correct the probability of rejecting the hypothesis 1/4 was .43. In particular, for the hypothesis 1/4 we have a probability of .05 that we will reject the hypothesis 1/4 even though it is true. (The probability that the hypothesis will be rejected if it is true is called the size of the critical region.)

5. The data used in paragraphs 3 and 4 were selected in a manner that tried to eliminate deafness by infection, i.e., by requiring two deaf children in the same family. Another method would be to use only those families where deafness occurred in more than one sibship. These data summarized from Table 4 are:

$N_2 = 10$	$N_5 = 12$	$N_8 = 2$	$N_{11} = 1$
$N_3 = 9$	$N_6 = 5$	$N_9 = 2$	$N_{12} = 3$
$N_4 = 7$	$N_7 = 6$	$N_{10} = 0$	
$M = 88$			

Using formulas 1 and 2

$$\overline{M} = 96 \qquad \sigma = 6.0$$

The critical region consists then of values of M outside the interval $96 \pm 2\sigma$ or 84 to 108. Again the observed value of M is not in the critical region, so we do not reject the hypothesis of probability 1/4.

6. We ask again the questions of paragraph 4, that is, if the actual probability is not 1/4 do we have a good chance of rejecting our hypothesis of 1/4? Calculation of these chances leads to the graph of the power function of the test.

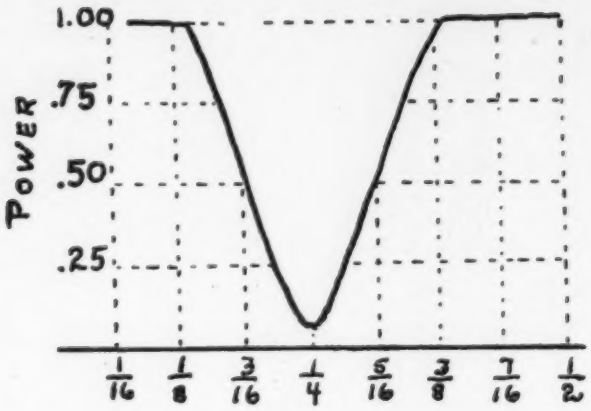


Fig. 2.

In this graph we note that the power curve is somewhat steeper near 1/4 than in the other case This means that the last experiment offers a better method of distinguishing between alternative hypotheses than does the preceding one.

A graph of the power function for the data in paragraph 1 gives

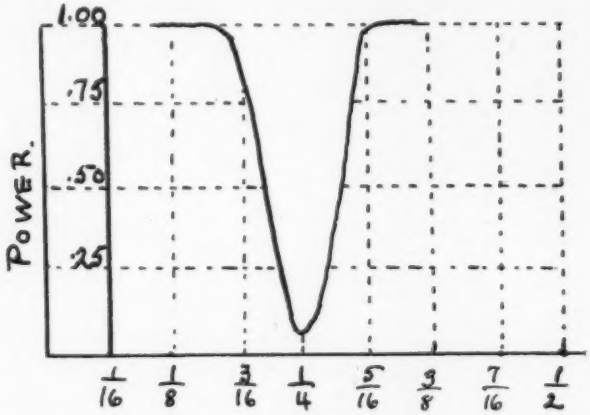


Fig. 3.

We note again that this curve is steeper than either of the other two, and hence would offer the best method of distinguishing between different hypotheses, provided that the data would not contain infectious cases.

7. Formulas to use in case the null hypothesis is that the probability

of the child being affected is p. Let $\overline{M}_2$ be the expected value of the number of affected children in N_2 families, $\overline{M}_3$ be the expected value of the number of children in N_3 families. And so on. Then mathematical calculation as in Hogben's Nature and Nurture, pages 75 and 124, shows that

$$5.\ \overline{M}_\kappa = \frac{kp}{1-q^\kappa} N_\kappa, \text{ where } q = 1 - p$$

The value $\overline{M}$ is obtained by adding the values of $\overline{M}_2$, $\overline{M}_3$, $\overline{M}_4$, ...

$$6.\ M = \overline{M}_2 + \overline{M}_3 + \overline{M}_4 + \ldots$$

Similarly for the standard deviations.

$$7.\ \sigma_\kappa^2 = \left(kp + q - \frac{kp}{1 - qk}\right)\overline{M}_\kappa$$

$$8.\ \sigma^2 = \sigma_2^2 + \sigma_3^2 + \sigma_4^2 + \ldots$$

Example. Suppose $p = 1/4$. Then $q = 3/4$.

Substituting in 5

$$\overline{M}_2 = \frac{2 \times 1/4}{1 - (3/4)^2} N_2 = 1.14N_2$$

$$\overline{M}_3 = \frac{3 \times 1/4}{1 - (3/4)^3} N_3 = 1.30N_3, \text{ etc.}$$

Using these values in 6

$$\overline{M} = 1.14N_2 + 1.30N_3 + \ldots$$

we get the formula 1 used in the first experiment.

In the case of the second experiment where only families of three children were used, the formulas for the expected value of the total number of affected children and the standard deviation computed by a method similar to Hogben's become:

$$9.\ \overline{M}_\kappa = \frac{kp\ (1 - q^{\kappa-1})}{1 - q^\kappa - kpq^{\kappa-1}} N_\kappa$$

$$10.\ \sigma\kappa^2 = \left(\frac{kp + q - q^{\kappa-1}}{1 - q^{\kappa-1}} - \frac{kp\ (1 - q^{\kappa-1})}{1 - q^\kappa - kpq^{\kappa-1}}\right) \overline{M}_\kappa$$

$$11.\ \overline{M} = \overline{M}_3 + \overline{M}_4 + \ldots$$

$$12.\ \sigma^2 = \sigma_3^2 + \sigma_4^2 + \ldots$$

THE USE OF X-RAY IN CHRONIC MASTOID CONDITION.

(a) CLINICAL ASPECTS.*†

HAROLD A. FLETCHER, M.D.,
San Rafael, Calif.

(b) X-RAY ASPECTS.

FRANK WINDHOLZ, M.D. (by invitation),
San Francisco Calif.

The problems with which the radiologist is confronted in cases of chronic mastoiditis are different from those in acute cases. This is due to the fact that in acute conditions the otologist is interested mainly with the anatomy of the mastoid and with pathological changes of the pneumatic system. Any additional information which he needs may be obtained from the history of the patient, from clinical examination and from otoscopic findings. Knowledge of the precise outlines and structure of the air cells and of the position of the sigmoid sinus and tegmen may influence the indications for and the planning of the operation and may also offer valuable points of view for prognostic considerations. The basic question in cases of acute changes is always: is a mastoiditis present or not? This question can be answered by the radiologist with more or less accuracy by a simple lateral view of the mastoid taken by Schuller or Law's position.

In chronic mastoiditis, however, the main interest of the otologist is concentrated on the middle ear and its accessory spaces: the mastoid antrum, attic and adjacent portions of the posterior wall of the external auditory canal. The basic question in these cases is not whether an inflammatory disease is present or not, because chronic mastoiditis always

*Read at the Seventy-eighth Annual Meeting of the American Otological Society, Inc., Chicago, May 31, 1946.
†From the Department of Surgery (Otolaryngology) and the Department of Medicine (Radiology), Stanford University School of Medicine, San Francisco, Calif.
Editor's Note: This ms. received in Laryngoscope Office and accepted for publication, July 23, 1946.

develops from a preceding acute mastoiditis. The otologist expects information regarding the presence or absence of a cholesteatoma and its precise description. It is of interest for him to know whether the attic and the posterior wall are involved in the disease, whether there are signs of perforations into the middle or posterior fossa, or recurrence of the inflammation.

The technique to be used by the radiologist to demonstrate the intricate anatomical parts involved in the disease varies according to the clinical considerations. Owing to the complicated anatomical structure of the middle ear and of adjacent parts of the mastoid, these areas are not as easily accessible to radiological exploration as are the cells of the pneumatic system.

TABLE 1.

Questions to Be Answered in Acute Mastoiditis	Technique
1. Development, extent, structure and precise outlines of pneumatic system	Schuller or Law's position
2. Presence of air cells in atypical localization	"
3. Structure of intercellular bone tissue	"
4. Air content of pneumatic cells	"
5. Signs of destructive bone changes	"
6. Signs of bone repair	"
7. Position of tegmen and sigmoid sinus	"
8. Additional questions according to the peculiarity of the individual case	According to actual problem

Tables 1 and 2 summarize the questions which the otologist expects to be answered by the radiologist in cases of acute and chronic mastoiditis. They also show which technique must be applied in order to demonstrate the anatomical structures involved.

Table 2 shows that in chronic mastoiditis the essential problems of the otologist cannot be solved without visualization of the mastoid antrum, attic, the external auditory canal and its posterior wall. To demonstrate these structures, an additional projection, namely, the axial view, is indispensable, because lateral views of the mastoid do not show these areas

with the clarity necessary for diagnostic purposes. This axial view was introduced approximately 20 years ago by Mayer, of Vienna, and only since that time can diagnostic

TABLE 2.

Questions to Be Answered in Chronic Mastoiditis	Technique
1. Development, extent, structure and precise outlines of pneumatic system	Schuller or Law's position
2. Presence of air cells in atypical localization, isolated cells	"
3. Structure of non-pneumatized bone	"
4. Condition of air cells	"
5. Signs of destructive bone changes in a) antrum b) attic c) posterior wall of external canal	Stanford or Mayer's position
6 Position of tegmen and sigmoid sinus	Schuller or Law's position
7. Changes of labyrinth	Stenver's position

TABLE 3. STANDARD RADIOLOGICAL VIEWS OF THE MASTOID AND ANATOMICAL STRUCTURES DEMONSTRATED.

Schuller or Law's position shows:
1. Pneumatic system
2. Bone structure
3. Position of sinus
4. Position and condition of tegmen
5. Position of jugular bulb

Stanford or Mayer's position shows:
1. Anterior and upper portions of pneumatic system
2. Anterior and posterior wall of external auditory canal
3. Mastoid antrum (dorsoventral view)
4. Relation of antrum to posterior wall
5. Attic (epitympanic recessus), lateral wall
6. Ossicles

Stenver's position shows:
1. Tip of the mastoid, including the cortex
2. Tegmen
3. Labyrinth
4. Internal auditory canal
5. Petrous apex

procedures be done on the middle ear and on its accessory spaces.

Table 3 enumerates the anatomical structures which can be demonstrated by using the techniques indicated.

The position successfully used by Mayer, however, presents technical difficulties which may account for the fact that it had not become popularized. It demands great skill of the technician and often personal attendance of the radiologist. Our attention has been directed towards simplifying Mayer's technique so as to make it accessible to the less skilled and less experienced technician. Impressed with this necessity, we constructed a chair in which the angulation of the skull is prearranged and the tube is in a fixed horizontal position, the beam directed perpendicularly in a plane connecting the outer margin of the orbit of the side not examined with the mastoid process being examined. The position of the mastoid process is given in a 45 degree rotation of the skull. This chair has been successfully used in the X-ray Department of Stanford Hospital. Continued improvements, which are still in progress have delayed its publication. The radiograph obtained in this position demonstrates the mastoid antrum, the walls of the external auditory canal and parts of the attic, as well as of the tympanic cavity, in a similar way as in the original position of Mayer.

It is hardly practicable to arrange the axial view in precisely the same angulation in all cases. This is due, not only to variations of technical data, but also to variations of the position of the mastoid process and petrous bones and to the spacial interrelation of these structures. In cases of changes of the projection the obtained radiograph can still be used because by application of the principles of parallax the normal appearance can be reconstructed. This is done by analysis of the positions of structures near the film and those near the tube. Even a minimal departure of the radiographic projection may induce considerable difficulties in interpretation of the radiograph, and an attempt must be made to correct these variations of the projection whenever they are seen. A dorsal deviation of the X-ray beam produces a ventral displacement of the petrous bone when related to the position of the mastoid, and vice versa. If the beam runs more parallel to the petrous bone, the latter is elongated. It is shortened in the reciprocal case.

Anatomical variations of the mastoid and petrous bones are very frequent. The size and height of the mastoid antrum varies between wide limits. The sigmoid sinus may overlie the mastoid antrum, being anteriorly displaced to a various extent. The width and height of the external auditory canal also shows considerable variations. All anatomical variations must be recognized in all its details. Their relationship to adjacent structures must be clearly established because an atypical appearance may lead to misinterpretations.

Simple chronic otitis in radiographs is, as a rule, associated with a more or less marked underdevelopment of the pneumatic system. Chronic suppuration is rare in cases of normal or approximately normal pneumatization. In such cases the inflammation is localized in the tympanic cavity and cells of the mastoid are not involved in the disease. In cases of underdevelopment of the pneumatic system, the non-pneumatized bone may be spongy or sclerotic. If only a few cells are present, they are irregularly distributed according to the underdevelopment of the pneumatic system in connection with the hyperplasia of the mucous membrane. In rare instances, pneumatic cells are distributed throughout the entire mastoid. If the inflammation extends from the tympanic cavity to the antrum and to the cells, the entire pneumatic system appears cloudy. It is of importance to know that this cloudiness may be due at times merely to the thickened hyperplastic mucous membrane and therefore may be only a result of disturbance of pneumatization and not of inflammatory changes. Thus, in cases of chronic otitis one is not justified in stating that an inflammatory condition is present in the cells when they are cloudy; however, if the antrum is clear, it is not likely that inflammation is established in the cells, excepting cases in which the otitis remains localized in the tympanic cavity. Bone changes do not develop in simple chronic otitis because the inflammatory changes remain localized in the hyperplastic mucous membrane and do not involve bone tissue.

The desquamative form of chronic otitis in its early stages seems no different from the simple form of chronic otitis. Here, too, one may find in the majority of cases

advanced underdevelopment of the pneumatic system. Concerning cloudiness of the antrum and of the cells, the considerations are similar to those in cases of simple chronic otitis. Consequently, from the radiological point of view there is only one absolutely definite sign of chronic inflammation, that is, the demonstration of a bone erosion.

Radiological signs of a bone erosion in the antrum in cases of chronic desquamative mastoiditis is the enlargement of the antrum in addition to its cloudiness. Borderline cases in such instances are not diagnostic because they may be considered as effects of a disturbance of pneumatization. If the arrest of pneumatization began only at the end of the first year, and a diminishing of the size of the antrum by subdivision of the lumen makes no further progress, the antrum remains large. Thus, a similar picture is obtained as it is seen when a small antrum becomes enlarged by bone erosion caused by inflammatory changes. Even serial examinations followed over several years do not give the possibility of differentiation because progress of the pathological process in the antrum is very slow.

Signs of bone changes in the surrounding of cells are also hard to establish if no serial examinations are at disposal. Cloudiness of cells, fuzziness of the outlines may be caused by inflammatory changes as well as by disturbance of pneumatization. If an inflammatory disease progresses from the cells into the diploe, it may remain for a longer time entirely unrecognizable.

Consequently, in order to make a diagnosis of bone changes these have to be somewhat advanced and obvious. They must be localized in a part of the mastoid easily accessible for X-ray examination and the appearance must be such as to be easily differentiated from that due to disturbances of pneumatization.

If the mastoid process is not completely sclerotic and cells are present, inflammatory changes are earlier recognized than if the sclerosis is complete. In such instances the outlines of the cells become fuzzy and bone trabeculae between

cells may disappear completely. Cells localized close to each other may then look like larger holes. If inflammatory changes involve the tegmen or the sinus plate, they are recognizable only if the margin of the pathological changes is located tangentially to the beam; otherwise they are not seen. At times the presence of varicosities of the sinus and an unusual anteposition may produce signs of bone destruction or even of enlargement of the mastoid antrum.

Bone destruction may be localized, not only in the antrum but also in the attic. Here, at times, there is difficulty in recognizing it because one sees the attic only in its craniocaudad axis and there is no way to demonstrate it in lateral views. In such cases the only X-ray sign we possess is markedly increased translucency indicating complete destruction of the lateral wall of the attic. Since, however, the height of the attic is variable, the density of the lateral wall is also variable. A bone destruction involving the lateral wall of the attic may be considered only when the Roentgen signs are obvious and a definite translucency is seen in radiographs. This involves the upper portion of the annulus tympanicus.

The ossicles are often not visualized in the axial view in normal positions. Their absence is of no diagnostic value and special projections are required for their demonstration. In most instances it can be disregarded of its demonstration.

In cholesteatoma suppuration, characteristic X-ray signs are seen. In early stages the only finding is a large, cloudy antrum with fuzzy, but regular, outlines. Air cells are less frequently present than in the exudative type of mastoiditis. If present, they are mostly in a condition similar to that of the antrum itself. With growth of the cholesteatoma, increasingly extensive bone changes are seen. These are localized sometimes in the antrum, at other times in the attic. If the cholesteatoma grows further, its margins become increasingly sharp. At the final stage one may observe a huge, nearly round translucency which involves the antrum and its surroundings. Extensions occur into the mastoid process and later into the upper portions of the posterior wall of the

external canal. On the posterior wall almost always the medial portion adjacent to the drum is affected. In further stages the bone which divides the antrum from the attic is destroyed and finally both spaces form a large hole which empties into the external auditory canal, destroying the upper portion of the tympanic ring. This condition is called nature's radical operation. In a characteristic way the anterior margin of the destruction forms a convex line pointing anteriorly and may be distinguished in such a way from defects after instrumental radical operation.

Occasionally nature's radical operation may develop from a cholesteatoma which does not originate in the antrum, but in the attic. In that event the primary changes are those of an attic cholesteatoma producing a rounded, punched-out translucency in the lateral wall and in adjacent parts of the tympanic cavity.

It should be mentioned that signs of an attic cholesteatoma can also be produced by an unusual height of the attic providing the lateral wall is thin.

As the inflammatory changes progress, the entire posterior portion of the external canal, and also the anterior wall, may be destroyed and in such a way again Roentgen signs of nature's radical operation are exhibited. The only difference is that the primary disease is localized in the attic, and not in the antrum.

The well known dense sclerotic margin of cholesteatoma develops only in late stages of the disease at the place where the matrix of the cholesteatoma is contiguous with the bone of the mastoid.

The Roentgen signs of a larger cholesteatoma may be summarized as follows:

1. A sharply outlined defect in the area of the lateral wall of the attic or of the antrum.

2. Faint increase of the density of the bone at the wall of the defect.

3. Absence of the upper portion of the posterior wall of the attic.

4. Confluence of the attic into the antrum, presenting the appearance of nature's radical operation.

5. Absence of ossicles.

In order to determine whether a cholesteatoma has perforated into the middle of the posterior fossa, Law or Schuller's position must be made. In these views medium-sized cholesteatomas become visible as faint translucencies in the upper lateral angle of the shadow of the petrous bone. With increase of the growth of the cholesteatoma the contour may be interrupted at some points, indicating that here the cholesteatoma has perforated; however, this is only visible when the borderline of the cholesteatoma lies in tangential position to the direction of the X-ray beam. At times absence of the shadow of a laterallly displaced sinus is suggestive of the presence of an extensive perforation into the posterior fossa.

Occasionally a cholesteatoma may destroy a part of the labyrinth. This condition is recognizable in Stenver's views, when the bony capsule of the lateral semicircular canal is destroyed and the canal itself communicates with the cavity of the cholesteatoma.

Acute recurrence of chronic otitis can only be considered in certain coincidence of circumstances. For instance, if the entire chronic inflammatory lesion is localized in the area of the tympanic cavity and all other parts of the mastoid appear to be normal. If in advanced stages of such conditions, acute inflammatory lesion develops in the pneumatic system, this becomes cloudy. This fact has to be considered as a recurrence of the disease. X-ray signs in such cases are identical with those of acute otitis.

Serial radiographs followed for a longer period of time may also disclose signs of an acute recurrence of the disease. These can be seen when the surrounding of an air cell, the appearance of which has not changed for a longer period of time, suddenly reveals abnormalities on its outline which were

not present previously. Most frequently we observe in such conditions translucent areas in the surrounding of such cells.

Finally, sudden interruption of the outlines of the wall of the cholesteatoma which did not change its appearance for a longer period of time has also to be considered in an acute recurrence of inflammatory changes.

Acute exacerbation may produce a perforation of the tegmen which is occasionally associated with brain abscesses.

Reparative changes following inflammatory lesions are characterized by increased density in the surroundings of previously established inflammatory lesions. In such cases it is important to know that disturbances of pneumatization may also produce sclerosis of the mastoid or of parts of it. Consequently bone sclerosis can be considered as a sign of reparative changes only if previous radiographs are present in which the sclerosis was not obvious.

Finally, following chronic inflammatory conditions, calcium deposits may be observed in the surrounding of the mastoid, which may be considered, at times, as calcified residues of inflammatory changes. Most frequently they are calcified thrombi of the sigmoid sinus, rarely calcified scars of an abscess of the temporal lobe. They have to be differentiated from calcifications occurring in the choroid plexus and in the pineal gland, the shadows of which are often projected into the surroundings of the petrous bone.

(b) X-RAY ASPECTS.*

FRANK WINDHOLZ, M.D.,
San Rafael, Calif.

The purpose of this paper, in the writing of which Dr. Windholz and I have collaborated, is particularly to bring out the importance of an X-ray projection in chronic mastoid conditions which has received very little attention in this country. This is the projection worked out by and named for Prof. Mayer, of Vienna. More specifically, it is to describe and demonstrate a simplified modification of the Mayer position which has been worked out at the Stanford Medical School and which we refer to as the Stanford technique. This projection, we feel, supplies a missing link in the use of the X-ray as an aid to diagnosis in chronic mastoid and middle ear conditions, and when understood will be much more widely used.

The fact that at least 25 different X-ray projections or positions have been described and used in the studies of mastoid conditions proves the difficulty to be overcome in obtaining all the information we want in one or two practical exposures. In the United States we have pretty generally narrowed the use of the X-ray down to two positions. These two are the Law or Schuller's position to show the normal or abnormal anatomy of the pneumatic system of the mastoid in acute and chronic conditions, and the Stenver position to show the normal and pathological condition of the petrous bone, the tegmen and capsule of the internal ear. We are quite familiar with these positions and it is not the purpose of this paper to review or supplant them. The purpose is to add a third position to our armamentarium to fill in the information lacking in Law and Stenver's position, particularly in chronic mastoid and chronic middle ear conditions.

In cases of chronic mastoiditis, where sclerosis obscures or

*Read by invitation at the Sixty-eighth Annual Meeting of the American Otological Society, Inc., Chicago, Ill., May 31, 1946.

completely blanks out any details of the area, neither the Law position nor the Stenver position gives much, if any, information as to the structure (normal or pathological) of the middle ear, attic, antrum, posterior wall of the external canal, or the meatus ad antrum. All of us at many times have had a feeling of frustration over not being able to get radiological help in this region. Some otologists have gone so far as to say that the X-ray gives us no help in such conditions. I have had many cases throughout the years that gave me a great deal of concern in deciding clinically whether they were properly to be considered surgical or non-surgical. Since we have perfected the use of this added projection in our X-ray studies, many of these cases have been much more clearly and correctly analyzed.

The projection or position necessary to bring out the details of these important areas is an oblique axial projection. Prof. Mayer provided this, but it was complicated and necessitated two angulations of the X-ray tube and three variations of the position of the head of the patient, and this complicated technique prevented the method from becoming widely used. The Stanford X-ray Department elaborated a simplified single axial view which brings out the desired structures.

In X-ray studies of the mastoid, it must be remembered that the demonstration of normal structures is often of as great importance as demonstration of abnormal structures, which may not be true in X-ray studies of other parts of the body. Our X-ray studies of chronic mastoid conditions now make use of three basic positions for visualizing the various parts of the mastoid. These positions are as follows:

1. For development, extent and structure and outline of the pneumatic system	Schuller or Law's position
2. For the presence of air cells in a typical localization—isolated cells	"
3. For structures of non-pneumatized bone	"
4. For condition of air cells	"
5. For signs of destructive bone changes in	
(a) antrum	
(b) attic	
(c) posterior wall of canal	Stanford technique

6. For position of tegmen	Stenver position
7. For changes of labyrinthine capsule	"
8. For position of sigmoid sinus and tegmen	Schuller or Law's position and Stenver's position

As previously stated, we are continually seeing cases of chronic or recurrent discharging ears and attic suppurations which puzzle us. Clinically, without waiting for complicating signs and symptoms, we are unable to differentiate the chronic middle ear and tubal infection (chronic or recurrent) from primary pathology in the attic and antrum, with perhaps potentially dangerous necrosis and cholesteatoma. Sometimes, erring on the side of conservatism, we defer operation until danger signs appear or intracranial pathology has insidiously crept in, or until permanent loss of hearing which might have been preserved has taken place. Many cases can be differentiated clinically and analyzed without the aid of radiology. There are those cases with a large dome-like attic opening which may be cleaned out or periodically spontaneously discharge a cholesteatoma, and which, if kept under reasonable observation, need no surgical intervention and are better off without. As Dr. Kenneth Day has pointed out in previous papers, a small dry attic perforation may not have a cholesteatomatous extension, and a moist attic perforation may be assumed to have a cholesteatomatous extension. It must also be remembered that cholesteatoma may not be constantly present in a given case and that a necrotic cavity of the attic, antrum or mastoid may be present without cholesteatoma at any time. The cavities which cause trouble, particularly from cholesteatoma, are those with a bottleneck outlet through a small attic perforation. If we can get radiological help in giving us further accurate information about an area which has previously been a blind area, we are in a much better position to decide on the type of procedure indicated. Use of the Stanford technique or Mayer position in a long series of cases has proved to us conclusively that this help is available.

One somewhat rare condition, the congenital absence of the external canal proved very interesting when studied with

this technique. The presence or absence of a pneumatized middle ear, attic and antrum may be visualized, which may have a direct bearing on the consideration of future operative procedures directed towards unlocking the obstruction of sound waves. We have three such cases under observation since birth.

One thing must be emphasized in the consideration of this method, and that is that cooperative study by the otologists and radiologists is of greater importance. The technique of taking the pictures by this method is not difficult. The interpretation of the film is difficult and will necessitate long and careful study of the part of both before it is mastered. From the otologist's standpoint, the projection is distorted and new because of the oblique planes involved, from the radiologist's standpoint because he must get down to detailed study of the normal and abnormal structure and development, with all its variations, of a very complicated area. Like a great many things in medicine which have proven worthwhile, mastery of them has come only through patient and long study.

GEORGETOWN UNIVERSITY POSTGRADUATE COURSE.

The Georgetown University Medical Center will sponsor an intensive graduate course in oto-rhino laryngology-bronchoscopy by Prof. Georges Portmann, of the University of Bordeaux, France. The course will begin April 7, 1947, and continue for two weeks in Georgetown Medical School, Washington, D. C. Address inquiries to: James A. Flynn, M.D., F.A.C.S., 1511 Rhode Island Avenue, N.W., Washington, D. C.

HEMOSTASIS AFTER ADENOID OPERATIONS.

ERNEST REEVES, M.D.,
Passaic, N. J.

"Postoperative bleeding from the adenoid is statistically more dangerous than from the tonsils." This sentence is quoted from one of our newest textbooks.[1]

This assertion seems to be at variance with the findings in previously printed statistics. Keen,[2] in 1931, found the percentage of postoperative adenoid and tonsil bleedings nearly the same. NcNally,[3] in 1927, found 7 per cent tonsil bleedings and only 4 per cent adenoid bleedings among his cases.

Richards,[4] in 1935, collected data of 10,475 cases with a percentage of serious after-bleeding for tonsillectomies 2.7 per cent, and for adenoidectomies 2 per cent. Jones,[5,6] in 1935, found 2.2 per cent tonsil bleedings and only 1.6 per cent adenoid bleedings in his 8,500 cases.

This seeming contradiction between our experiences of today and the experiences of only 10 to 15 years earlier is probably caused by the fact that today hardly a specialist would allow his patient to leave the operating table without stopping every evidence of bleeding from the tonsillar fossa. Although many surgeons are just as careful in stopping the bleeding in the adenoid fossa, this method of operation is not so generally accepted as is the hemostasis after tonsillectomies.

There are few events in the daily life of the laryngologist that cause more annoyance than the occasional telephone call three to six hours following an uncomplicated T and A operation, announcing that bright blood is trickling from the nostril, or that the child has been vomiting blood in appreciable quantity. Not the least unpleasant aspect of the situation is the necessity to explain to the parents as to why the bleeding

Editor's Note: This ms. received in Laryngoscope Office and accepted for publication, Sept. 24, 1946.

has occurred, or why it is necessary to return the child to the operating room.

There may be operators who can look over a long and busy career and fail to recall any instance of alarming postoperative adenoid bleeding. For those men, who are responsible for a conduct of a hospital clinic, in which the work necessarily has to be done by a changing and a less experienced staff, there will inevitably occur a certain number of cases of postoperative bleeding requiring prompt control.

It is perhaps unfortunate that the adenoidectomies are often regarded and treated as an appendix to tonsillectomies.

In most articles and books, only one method, *i.e.*, the retronasal packing, is extensively described. The other equally important ways of hemostasis are usually only perfunctorily mentioned.

This article tries to describe the chief procedures which help us to get a dry field after adenoidectomies just as we have after perfect tonsillectomies. It is, of course, quite obvious that a major part of the postoperative bleeding could be avoided by careful examination and selection of the patients.

The operation is contraindicated in hemophilia. A postponement is indicated in the several forms of purpura, in the presence of acute infectious diseases, especially influenza, measles, scarlet fever and agranulocytosis; in fact, in any case of elevated temperature. It is better to avoid adenoidectomies during the menstrual period, secondary anemia, or during active treatment by arsenic or bismuth in syphilis.

Preoperative testing of the bleeding time, coagulation time, and in suspicious cases the counting of platelets is advisable.

Most specialists use five methods of hemostasis after tonsillectomies — the plain ligature, the suture ligature, packing, electrocoagulation and infiltration of the tissues with novocain-adrenalin. As to which one of these procedures is used depends upon the individuality of the surgeon, the way he wants to perform the operation, the cause of the bleeding and, most important, upon the anatomy or pathology of the

bleeding site. Many laryngologists dislike the suture ligature on the lateral wall of the tonsillar fossa, because during the act of suturing the needle disappears from view and, as we know from the anatomy, it may injure the important vessels and nerves, or may carry infections to the parapharyngeal tissues. Even more important in the selection of the most appropriate way of hemostastis is the topographical anatomy of the adenoid. It is situated in a place which is very much more difficult of access, and where the result of any injury to the neighboring organs may become very deleterious; therefore, we try to delineate that part of the applied anatomy of the pharynx which is important for our present purpose.

Although many authors consider Luschka's "Pharynx of the Man"[7] the first really useful dissertation of the adenoid region, we find in the literature many other excellent treatises about this part of the human body before and after the appearance of Luschka's book which was published in 1860.[8-14] Exceptionally brilliant descriptions of this region were published by Testut[15] and lately by Barnhill.[16]

The rhinopharynx for our purposes could be considered as a box with six sides. The anterior side is always open in healthy human beings. This is the communication with the nasal cavity through the choanae. The inferior wall is formed only during the act of swallowing and for the formation of certain sounds. In adults, the capacity of this box is about 14 cc. It is about 2 cm. deep, 1.8 cm. high and 3.5 cm. in breadth.

The fornix begins at the nasal septum. If after elevating the palate, we look at the rhinopharynx, we would find it difficult to see a distinct margin separating the fornix from the posterior wall. It looks like and is a continuous curved surface.

According to Testut[15] there are three kinds of rhinopharynges. The most frequent type is the nasopharynx "a voute ogivale." In this form the top of the pharynx is high. In one-third of the cases we may find what Testut calls nasopharynx "a voute cintre," where the fornix is more rounded

and not as high as in the first type. For our purpose here, the most important type is the third type, "voute surbaissee." This is the infantile type. This type is low, the fornix lies in the height of the lower turbinate, and the tubal ostium is on the height of the palate. Because it is so low, the surgeon can more easily apply clamps and ties than onto the first type.

The fornix is firmly anchored to the sphenoid, the base of the occipital bone, and to the connective tissue filling out part of the foramen lacerum. The superior wall of the nasopharynx often continues without any visible landmark into the choanae, and at other times a fold or furrow may separate the two structures.

The vertical length of the posterior wall is 1.5 cm. If we penetrate the soft tissues in the midline, we find behind them the posterior part of the body of the occiput which is covered by a thick, fibrous layer, and a little lower down a ligament about 6 mm. thick, connecting the anterior magin of the foramen magnum and the anterior arch of the atlas. Behind the lower border of the rhinopharynx in the midline we find the body of the second vertebra. On both sides there is soft tissue made up of the longus capitis and anterior rectus muscles.

The anterior posterior diameter of the lateral wall of the rhinopharynx is about 1.5 cm. The most prominent feature of this wall is the Eustachian tube. It resembles a finger pointing from backward to forward and at the same time inward and downward. In that way the lateral wall of the pharynx is pushed inward, mostly by the medial part of the tube. The posterior wall of the tube is, therefore, covered about 1 cm. of its length by the pharyngeal mucosa. It forms a pocket, the pharyngeal recess or Rosenmüller fossa. The anterior wall of the recess is the cartilaginous tube, and the posterior wall of this recess is the most lateral part of the posterior wall of the pharynx. The lateral wall of the recess is strengthened by the lateral pharyngeal ligament and is in contact with the parapharyngeal space. Downward, this recess is often closed by a fold of mucous membrane, but sometimes extends smoothly over into the groove formed by the lateral and posterior walls.

The extent of this pocket varies, depending upon the amount of lymphoid tissue present. This sometimes fills out the whole recess and covers the pharyngeal end of the tube. The Rosenmüller fossa is not very strongly accentuated in children. It seems to have been developed from the second branchial cleft. If it is abnormally developed, it forms a diverticulum called the diverticulum of Pertick.[15]

From the lower end of the tube arises a prominent fold, the salpingopharyngeal plica. This is saturated with glands and very often is infiltrated by masses of lymphoid tissue. Sometimes it extends deep down and is visible below the soft palate on the lateral wall.

From the anterior tip of the mouth of the tube originates a short vertical fold, the salpingopalatine plica, which helps to form the lower half of the lateral side of the choanae and ends in the mucosa of the palate.

The pharynx is composed of three coats: an external fibrous layer which is very loose and allows the necessary movement of the pharynx, a muscular coat and the mucosa. The deeper parts of the submucosa form a special strong layer, the pharyngeal aponeurosis. In the upper part of the pharynx, the muscular coat is absent, and this aponeurosis unites with the external fibrous coat and forms a very strong tissue connecting the pharynx with the base of the skull.

From the muscular coat only the superior constrictor muscle is important for the purpose of this article. It arises from the lower third of the posterior margin of the medial pterygoid plate, from the pterygomandibular raphe, from the alveolar process of the mandible, and by a few fibres from the side of the tongue. The fibres curve backwards and are inserted into the median raphe in the posterior wall of the pharynx. The superior fibres arch beneath the levator veli and the auditory tube. There is no muscle in the upper part of the pharynx, only the aponeurosis pharynx, which here is called "sinus Morgagni." For us it is important to know that there is usually no muscle in the pharyngeal wall behind the adenoid.

The levator veli palati arises from the undersurface of the apex of the petrous part of the temporal bone and from the medial lamina of the cartilage of the auditory tube. After passing above the upper concave margin of the superior pharyngeal constrictor, it spreads out in the palatine velum. It causes an impression on the lateral pharyngeal wall which is often visible.

The only other muscle which is of interest in this applied anatomical description is the salpingopharyngeal muscle. It arises with a short, flat tendon from the lower thick part of the cartilaginous tube and passes downward close to the mucosa. When using suture ligatures in this region these last two described muscles should be remembered. Injury to these muscles may disturb the functioning of the tube.

Except for the soft palate which is covered by squamous epithelium, the mucosa of the rhinopharynx is mostly covered by ciliated epithelium. In it lodges the pharyngeal tonsil. It may be considered a lymphoid infiltration of the lamina propria of the mucosa. It consists of several folds with deep furrows between them. In the middle of the adenoid the furrows run nearly sagitally. On the side of the organ the furrows form arches which converge anteriorly and posteriorly.

Besides these furrows, the pharyngeal tonsils are perforated by numerous ducts which are covered with epithelium. These ducts do not end blindly as the crypts of the palatine tonsil, but are connected with the very numerous glands which lie behind the adenoids.

The so-called tubal tonsil is a collection of lymphoid tissue on the tubal torus. Lymph nodes are rarely formed here, but the infiltrated mucosa is disrupted in many places by small holes. These are the openings of the excretory ducts of the glands and these give the impression of tonsillar tissue to the observer.

Three arteries convey blood to the pharyngeal tonsils. The most important is the ascending pharyngeal artery. It arises from the external carotid and ascends vertically between the internal carotid and the pharynx to the base of the skull.

Three pharyngeal branches originate from it to the three parts of the pharynx. The upper branch, after arriving at the base of the skull, turns downward and this is the chief supply of blood for the rhinopharynx.

Two smaller arteries are worth mentioning: the supreme pharyngeal wall, which is a branch of the sphenopalatine artery, and the Vidian artery, a branch of the descending palatine artery. It passes backward along the pterygoid canal with the corresponding nerve. It sends branches to the upper part of the pharynx and the auditory tube.

There is an abundance of veins in the adenoids. In the newborn they make up one-third of the mass of the pharyngeal tonsil.[14,15] These veins are the most common sources of bleeding after an adenoidectomy. The blood of the venous pharyngeal plexus is conveyed into two veins. One goes upward and empties near the base of the skull into the internal jugular vein. The other usually goes downward.

Many authors accentuate the necessity of removing every loose tag of tissue.[17] This seems to be the primary requirement to prevent excessive postoperative hemorrhage. The best and easiest way to achieve a clean operative field is to perform at least the end of the operation under direct visualization.[18,19] Some surgeons constructed special instruments for that purpose.[20,21]

We usually use the method described by Dr. Fetterolf,[22] with slight modifications. The tonsils are removed first. No packing or other kinds of hemostasis is done in the tonsillar fossa in this stage of the operation. As soon as the tonsillectomy is done, a LaForce adenotome is introduced high up and the central portion of the adenoid is removed. If we feel that the adenotome did not remove all the tissue, we use a curette afterwards. At this time a Tieman elevator is introduced, pushing the palate out of the way. This gives an excellent view of the fornix in that type of low, flat pharynx, prevalent among children, which is called by Testus "voute surbaisse."

We clear the field with suction, and we have a clear view of the loose tags, remnant of adenoid tissue, and the thick,

infiltrated mucosa of the posterior wall. The removal of all the visible lymphoid tissue is easily executed by a punch of the Myles type.

The strongly infiltrated salpingopharyngeal plica can now be seen as a thick, ragged curtain hanging down behind the posterior pillar. In removing this tissue the surgeon should not go too deeply, to avoid injuring the muscle behind the plica.

In this stage no packing is left in the adenoid fossa; instead we turn now our attention back to the tonsil and carefully stop every oozing site. This usually takes a few minutes. During this time the adenoid, in most cases, stops bleeding. According to Lyman Richards[4] the usual bleeding time after an adenoidectomy is only four to six minutes.

We now elevate the soft palate and examine the adenoid region. Many authors accentuate the fact that the disturbing of the clot causes the fossa to bleed anew; therefore, the coagulated blood is first removed cautiously with a forceps. It is not absolutely necessary to remove all the clots from that part of the fornix which is near the nasal cavity. Most of the bleeding comes from the abundant venous supply of the mucosa. If all the mucosal tags were removed from the fornix during the operation there would very rarely be any serious bleeding from there. The bleeding comes usually from the edges of the pharyngeal mucosa below the extirpated adenoid. The slight amount of blood remaining after the removal of the firm clots by the forceps is now skimmed away from the edge of the mucosa so that we can see the whitish surface of the pharyngeal aponeurosis. The edge of the mucosa is rarely a clean-cut horizontal line. The curette and punch cut pieces out of it so that the edge is indented and looks like one or several W's lying near each other. Often it bleeds from two or three points. The point which seems to bleed the most is grasped by a six to seven-inch Allis clamp with four mouse teeth.

The modified Coakley[23] slipknot is now applied. Although this modification is used by many surgeons, it makes hemo-

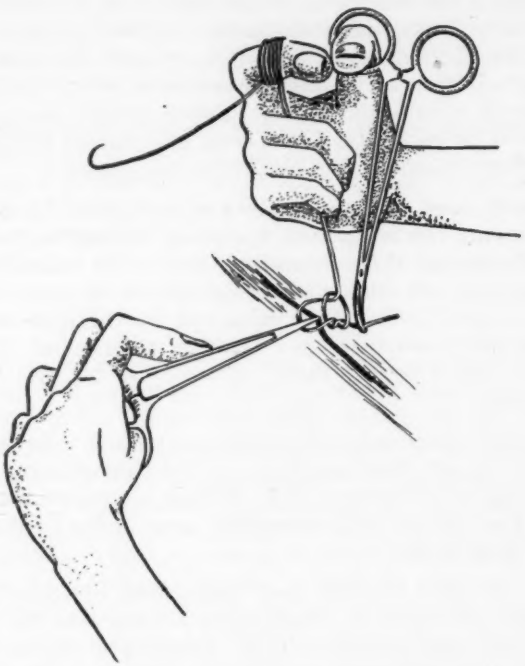
stasis in the adenoid fossa so much easier than the original Coakley slipknot that we describe it here.

The difficulty of operating in a deep hole through a narrow mouth, as in a tonsillectomy, is very much enhanced in adenoid bleeding. The surgeon and his assistant cannot bring their heads in such a position that they should both be able to observe the operative field at the same time. If the assistant handles the Allis clamp, he does this blindly. This makes tying a very uncertain procedure and often leads to failure. The modification allows the surgeon to tighten the tie with his right hand only. His left hand is free to juggle the Allis clamp in the right position and to remove it at the precise moment.

The slipknot holds the ligature only by friction. If the friction is not strong enough, the knot will not hold; conversely, if the friction is too strong, the catgut will not slip at all and the knot cannot be tightened.

Every surgeon has his individual preference in catgut. Davies and Geck's non-boilable No. 1 plain catgut is used widely. The forming of the knot is described excellently by Coakley,[23] and later by Moore;[24] therefore, we delineate only those parts of the method which Coakley did not mention in his article. Preliminary testing will show us whether the friction is satisfactory. If at the trial we feel that the knot does not hold tightly, we put the catgut in sterile water for one or two seconds. This enhances the friction. If we are satisfied that the knot is correct, the short end is grasped by long-handled forceps close to the knot. The end of the forceps should be slightly curved and serrated. The loop is now passed over the handle of the Allis clamp, which has grasped the bleeding point. By pulling the long end of the ligature the loop is narrowed to one-third of an inch. The thumb of the right hand is hooked into one of the rings of the long-handled forceps, and the long end of the catgut is fastened by three or four turns onto the second finger of the right hand. To be surer, the end of the catgut could be fastened by a knot to the fourth finger, or it can be pressed between the lower surface of the right thumb and the ring of the long-

handled forceps. The length of catgut between the fourth finger of the right hand and the knot should have a slight slack. In this slack the third, fourth and fifth fingers of the right hand should be inserted. If we move these fingers away from the forceps the knot will be tightened.



The Allis clamp is now seized in the left hand of the surgeon, who manipulates it so that the knot will tighten the tissues below the jaws of the clamp. If there is any difficulty, the elevation of one end of the Allis clamp or a half turn helps to slip the catgut below the clamp. The mucosa is pretty

loosely attached here to the submucosa and can be moved relatively easily.

After the cautious removal of the Allis clamp, the third, fourth and fifth fingers of the right hand tighten the knot thoroughly. This tightening is necessary because the tissue beneath the Allis clamp is just as flat as the grasping end of the clamp. The tightening of the knot, after removal of the Allis clamp, forms this tissue into a fold with a tight waist and a broad, loose end. This fold prevents the knot from slipping. During all of these procedures, which really last only about a half-minute, the surgeon should have a clear field, done by suctioning blood from the edge of the mucosa, just before he applies the catgut on the bleeder.

In most cases only one ligature is necessary. If the most active oozing bleeder is tied, the other smaller bleeders will stop. The causes of the hemostasis may be the added time, or the elevating and folding of the mucosa in the first tie may cause a slight torsion of the veins and may even be covering some of the previously open bleeding points; but the fact remains that it is very rarely necessary to put two ties on the mucosa.

The above described method of hemostasis is very easy and satisfactory, but there are places on the rhinopharynx where the placing of a ligature will be difficult, as the cartilaginous part of the tube, or the Rosenmüller fossa, or the tight tissues on the aponeurosis.

In these areas the best and easiest way of stopping the bleeding is the injection of adrenalin solution into the tissues around the oozing points. Grant[25] describes a similar procedure. It is, of course, well known that if we want to perform an adenoidectomy on an adult with local anesthesia, the injection of the posterior wall of the rhinopharynx with novocain-adrenalin solution not only makes the operation painless but simultaneously also stops the bleeding.

The injection should be made with a thin, strong needle, as the one used for local anesthesia in tonsillectomies. The wall of the pharynx is relatively thin. The needle should be

parallel to the long axis of the pharynx to avoid injuring the big vessels or nerves. After the injection of 5 to 10 cc. of this solution the bleeding stops. The adrenalin keeps the vessels contracted for one to one and one-half hours, in which time normal blood will clot.

In this stage of the operation the sandbag should be taken from below the shoulders, the extended head should be brought into the normal position and the mouth should be closed. The abnormal position of the neck and jaw will cause torsion and stasis in the neck veins and so promote hemorrhage.

If these methods are not possible or are not satisfactory, we may use the third method, the retronasal packing. Although this way of hemostasis after an adenoidectomy is the one which is most commonly used and described, it has several disadvantages. It makes the patient very miseraable. Children often refuse to take even water. It stops the drainage and ventilation in the nose, tube and rhinopharynx, and so promotes the development of an infection.

If there is profuse oozing from many sites as in hemophilia, purpura or other diseases causing intractable bleeding, we have to resort to the retronasal packing, but if possible, it should not be done blindly. We should examine under direct vision the source of the bleeding and place the packing so that it exerts its pressure on the bleeding points. The packing should be solid, compact and of sufficient size to prevent slipping into the choanae. This latter event is useful and desirable in cases where the retronasal packing is used to stop an otherwise intractable epistaxis by closing the choanae and sealing the nasal cavity posteriorly, but in bleeding of the rhinopharynx this is not necessary. Conversely, a packing pulled into the choanae may not exert sufficient pressure on that part of the posterior wall of the pharynx which oozes.

The packing is pulled to the bleeding site by a small sized rubber catheter which is introduced through one of the nostrils and is brought out through the mouth. The packing should be made compact with a piece of strong, black silk. To prevent the string slipping around the packing, we push a strong nee-

dle threaded with black silk through the packing and knot the silk on that part of the gauze packing where we want it, usually in the middle of the gauze. The silk is fastened to the catheter and is pulled through the nose, and the two ends are caught by a hemostat. The palate should be elevated and the packing should be placed with the help of a curved hemostat so that it presses on the bleeding point. The silk hanging from the nostril should not pull the packing into the choanae. Its only purpose is to fix the packing so that it does not slip down. If the packing is placed well on the oozing part of the rhinopharynx, packing of the nose is not necessary.

The fourth method of hemostasis is the suture ligature which gives excellent results, but it has several serious disadvantages. The forming of a tight knot in the narrow, deep hole is not easy. The surgeon must be careful not to injure the levator veli and salpingopharyngeal muscles, because they are important for the functioning of the Eustachian tube. The needle should not go too deeply, to avoid carrying infection into the parapharyngeal space.

The last method used for the stopping of oozing is electrocoagulation. It is usually used in conjunction with the removal of individual granulations and lymph nodules in the posterior pharyngeal wall. It may be used either unipolarly, by touching the hemostat which grasps the bleeding point with the electrode, or by applying the two-pronged electrode. With these instruments the surgeon coagulates the tissue beneath the hemostat.

After even the most careful operation some bleeding may start either a few hours or a few days later.

The cause of this bleeding is not easy to ascertain in every case. Vomiting may push the ligature down, or a sudden rise in blood pressure may eject a loose clot. Local hyperemia may be the first signs of measles, scarlet fever, influenza, etc., causing capillary oozing of the cut surfaces. The sloughing of loosely hanging, badly nourished tags may open new blood channels. Easily bleeding granulations may be injured by swallowing or by a misplaced bolus. It seems that a vitamin C

deficiency promotes these after-bleedings, as does the use of salicylates, etc.[26]

In most cases the bleeding is easily stopped by dropping adrenalin solution into the nostrils. The best method is to repeat the drops every few minutes until we are sure that the nasal cavity is dilated by the adrenalin, and permeable, so that the drops are able to reach the bleeding surface of the rhinopharynx. Then two drops every two hours usually will keep the oozing in check. If there is a clot hanging down into the throat it should be removed with forceps. The clot irritates and excites the patient, and prevents the adrenalin from coming in contact with the source of the hemorrhage.

At the same time, vitamin K, thromboplastin or calcium gluconate may be supplied to promote hemostasis. Some men use extract of pituitary.[27] Vitamin C should be given either in the form of orange juice or ascorbic acid before and after operation. Salicylates should not be used extensively.[26]

A transfusion is highly recommended by many surgeons. It serves not only to replace the blood, but at the same time contains all the ingredients present in normal blood which are necessary for coagulation.

In the very rare cases in which these methods do not suffice to stop the bleeding, one of the methods described above has to be used. If the surgeon is able to see the bleeding site, injection of adrenalin solution is the easiest and most satisfactory way.

SUMMARY.

1. The bleeding after an adenoidectomy should be as carefully stopped as the hemorrhage after a tonsillectomy.

2. The anatomy of the rhinopharynx is described to evaluate the best method of hemostasis in a given case.

3. The most frequent source of hemorrhage is the cut edge of the pharyngeal mucosa below the adenoid, where the bleeding is easily stopped by the modified Coakley's slipknot ligature.

4. The second best method is the injection of adrenalin solution.

5. In cases where a retronasal packing has to be used, it should be so placed that it exerts pressure on the bleeding surface.

REFERENCES.

1. JACKSON and JACKSON: Diseases of the Nose, Throat and Ear, p. 163. 1945.

2. KEEN, J. A: Abnormal Hemorrhage After Tonsil and Adenoid Operation. *Jour. Laryngol. and Otol.*, 46:297, 1931.

3. MCNALLY, W. J.: Hemorrhage Following the Removal of Tonsils and Adenoids. *Canad. Med. Jour.*, 17:690, 1927.

4. RICHARDS, LYMAN: Some Considerations of Adenoid Bleeding. *Ann. Otol., Rhinol. and Laryngol.*, 44:117, Mar., 1935.

5. JONES, DAVID H.: Observations and Result in Adenoid and Tonsil Surgery. THE LARYNGOSCOPE, 40:667, Sept., 1930.

6. JONES, DAVID H.: The Administration of a Tonsil Clinic in a Nose and Throat Hospital. *Bull. Am. Col. of Surg.*, 17:1:35, Mar., 1933.

7. LUSCHKA: *Schlundkopf des Menschen*, 1868.

8. CRUVEILHIER, J. : Descriptive Anatomy. London, 1842.

9. BAKER, M. J.: Waldeyer Ring. *Ky. Med. Jour.*, 37:152-156.

10. STURM, F. PIERCE: Anatomy of Adenoid. *Practitioner.*, 129:560, Nov., 1932.

11. SOBOTTA: *Hanb. der Speziellen Chir. des Ohres u. Oberen. Luftwege*, B. 1, 1913.

12. IMHOFFER, R.: *Handb. der Speziellen Chir. des Ohres u. Oberen. Luftwege*, B. 2, p. 223.

13. WETZEL, G.: Anatomie und Entwickelungsgeschichte der Mundhohle und Raches. *Handb. der Hals., Nasen u. Ohrenheilk.* Denker and Kahler, VI.

14. SCHUMACHER, S.: Histologie der Luftwege und Mundhohle. *Handb. der Hals., Nasen u. Ohrenheilk.*, Denker and Kahler, VI.

15. TESTUT, L.: Traite D'Anatomie Humaine. Huitieme ed., p. 128, G. Doin Co., Paris, 1931.

16. BARNHILL, JOHN F.: Surgical Anatomy of Head and Neck. 1937.

17. HILL, FREDERICK T.: Observation Following Adenoidectomy. *Arch. Otolaryngol.*, 14:775, Dec., 1931.

18. HUBBY, L. M.: Some Remarks on Tonsillectomies and Adenoidectomies. *Med. Rec.*, 99:641, 1921.

19. Beck, J. C.: Removal of Adenoids by Direct Inspection. *Ann. Otol., Rhinol. and Laryngol.*, 22:273, 1913.

20. Kelley, I. D., Jr.: Adenoid Removed with Direct Vision Adenotome. *Ann. Otol., Rhinol. and Laryngol.*, 32:1053, 1923.

21. Guggenheim, Louis: Direct Adenoidectomy. *Arch. Otolaryngol.*, 33:314, Feb., 1941.

22. Fetterolf, George: Abstract of Discussion, Eighty-second Annual Session of the American Medical Association, Philadelphia, June 10, 1941. *Arch. Otolaryngol.*, 14:779, Dec., 1931.

23. Coakley, C. G.: Hemorrhage During and After Tonsillectomy. *Jour. Laryngol. and Otol.*, 36:3, 1922.

24. Moore, Irvin: The Tonsils and Adenoid and Their Diseases. London, 1928.

25. Grant, Heman: Hydrostatic Pressure in Adenoid Hemorrhage. *Ann. Otol., Rhinol. and Laryngol.*, 53:576, Sept., 1944.

26. Neivert, Harry; Pirk, Leo A., and Engelberg, Recha: Late Sec ondary Hemorrhage. *Arch Otolaryngol.*, 43:568, June, 1946.

27. McLaurin, J. W.: Use of Solution of Pituitary of Twice U. S. P. Concentration for Hemorrhage Following Tonsillectomy or Adenoidectomies. *Arch. Otolaryngol.*, June, 1944.

J. BISHOP ANNOUNCES FIRST HYPONEEDLE WITH PLASTIC HUB.

The first postwar improvement in hypodermic needles, a needle with the first plastic hub, was announced by Paul C. Kerk, president of J. Bishop and Co., Platinum Works. The hub of the Albalon needle, as the company calls this innovation, is made of gleaming white plastic nylon. The needle itself is of stainless steel and is so beveled as to cleanly pierce and spread the epidermis without undue cutting, slicing or bruising the skin. This also provides a less painful injection.

The plastic hub withstands all commonly used methods of sterilizing, eliminates freezing of hub and syringe tip and thus tends to reduce breakage of syringe from this cause. Leakage around syringe tip is also minimized by the elastic qualities of the Albalon hub. The company has just released the Albalon needle for distribution through the usual trade channels, and will promote the sales of this and the Bishop production in the leading medical and hospital journals.

THOMAS E. CARMODY, M.D.

IN MEMORIAM

THOMAS E. CARMODY, M.D.,

1875 - 1946.

Dr. Thomas E. Carmondy died while asleep on a Branif plane flying from Denver to Pueblo, on Friday afternoon, Aug. 30, to attend a dinner in honor of Dr. Wm. H. Baker, a very prominent Pueblo general surgeon. When the stewardess attempted to awaken Dr. Carmody, she found him dead.

Dr. Thomas Edward Carmody was born in Shiawassee County, Mich., May 22, 1875. He was a son of Thomas and Mary Ann Carmody of Irish parentage. His early life was spent in Owasso, Mich., where he graduated from high school. He attended the University of Michigan Dental School, where he received a D.D.S. degree in 1897. In 1908, he received a D.S.Sc. degree from the same school.

He located in Denver in 1897 for the practice of dentistry and attended the Denver and Gross College of Medicine, where he received an M.D. degree in 1903. He was professor of bacteriology and histology at the University Dental College from 1898 to 1905, and professor of oral surgery and rhinology at the same dental college from 1905 to 1932. He was assistant in laryngology and otology at the Denver and Gross College of Medicine from 1905 to 1911. On Jan. 1, 1911, the University of Colorado absorbed the Denver and Gross College of Medicine and the latter passed out of existence. He continued with the Department of Otolaryngology until 1912 at the University of Colorado School of Medicine in Denver. He was connected with the Research Department of the Child Research Council, as an otolaryngologist, at the University of Colorado School of Medicine from 1928 to 1936. The Child Research Council is in the Department of Pediatrics.

Dr. T. E. Carmody became a Fellow of the American College of Surgeons in 1913, of the American College of Dentists

in 1929, and of the International College of Surgeons in 1941.

He became a member of the Denver Dental Society in 1897 and was its president in 1907. He became a member of the Denver City and County Medical Society in 1903; was its secretary in 1904 and president in 1923. He joined the Denver Clinical and Pathological Society in 1912 and was its president in 1931. He became a member of the Colorado State Dental Association in 1897, and of the Colorado State Medical Society in 1903. He was the first president of the Colorado Society for Crippled Children. The Colorado Otolaryngological Society was founded about 1910 and held meetings for about five years. It was reorganized in 1918 and 1919, at which time he became the first president of the reorganized society.

During World War I, Dr. T. E. Carmody was commissioned a major, M.C., U.S.A., and was stationed at Fitzsimmons Army Hospital. He had held the rank of first lieutenant in the Medical Corps from 1914 to 1918.

He was a charter member of the American Board of Otolaryngology and joined the American Academy of Ophthalmology and Otolaryngology in 1903. He was president of the Academy in 1923. He belonged to the American Bronchoesophagological Society and was its president in 1930. He joined the American Laryngological, Rhinological and Otological Society in 1912 and was its president in 1936. He joined the American Laryngological Association in 1924 and was its president in 1941. He became a member of the American Otological Society in 1925. He became a member of the A.M.A. early and was chairman of the Ear, Nose and Throat section in 1931. He was a member of the American Society of Oral and Plastic Surgeons.

He held honorary memberships in the Pacific Coast Oto-Ophthalmological Society, the Texas Ophthalmological and Otological Society, the Kansas City Ear, Nose and Throat Society and the Illinois State Medical Society. He was a mem-

ber of the first International Otolaryngological Congress in Copenhagen, Denmark, 1929. He was a member of the Board of Directors of the National Society for Crippled Children. He took postgraduate work, as an assistant to Dr. T. W. Brophy and in American clinics.

He held staff appointments as oral and plastic surgeon at Children's Hospital, Denver; consulting otolaryngologist and bronchoesophagologist, St. Luke's and Mercy Hospitals, Denver; Evangelical Lutheran Sanitorium, Wheatridge; Swedish National Sanitorium, Englewood; and Jewish Consumptive Relief Sanitorium, Spivak.

He was a contributor on plastic surgery to the Jackson, Coates and Jackson textbook on ear, nose and throat diseases in 1929, and on diseases of the mouth and tongue for the Jackson and Jackson book in 1945. He wrote numerous articles covering diseases of the mouth, nose, throat and ears. He was a member of the editorial board of the *Annals of Otology, Rhinology and Laryngology.*

On Nov. 7, 1899, he was married to Mary Jane McBride in Denver. Mrs. Ruth Carmody Sumners, Denver; Mrs. Mary Alice Carmody Cobb, Minneapolis; and District Attorney David W. Carmody, Santa Fe, N. M., are the surviving children.

His hobbies were golf and photography. He belonged to the Denver and University Social Clubs and the Denver Country Club. He was an Episcopalian.

Dr. T. E. Carmody was very devoted to his wife and children. His wife accompanied him to all board and medical meetings and took an active interest in his professional work. Denver and Colorado lost a distinguished otolaryngologist, a great citizen of the Rocky Mountain region, and a loyal member of the medical and dental professions. F. R. S.

WELLS P. EAGLETON, M.D.

WELLS P. EAGLETON, M.D.,

1865 - 1946.

It is with the deepest regret that we record the death of Dr. Wells P. Eagleton, of Newark, N. J., on Sept. 12, 1946, at Portsmouth, N. H., where he had spent the summer.

Dr. Eagleton was born in Brooklyn, N. Y. He received his M.D. degree from Columbia College of Physicians and Surgeons in 1888, after which he interned at City Hospital, Newark, N. J., for two years before establishing an office for private practice. He founded the Newark Eye and Ear Infirmary and served as its medical director, chief of staff and superintendent.

His fifty-six years of practice in Newark were interrupted only during World War I when he served as Chief of Brain Surgery at Camp Dix Base Hospital.

In 1922, he published his book, "Brain Abscess, Its Surgical Pathology and Operative Technique." It soon became a text in medical colleges and was the beginning of numerous volumes in his special field of medicine. Later, his books, "Cavernous Sinus" and "Thrombosis," together with numerous monographs, reflected his important research in these and kindred subjects and placed him high in the esteem of the profession.

Many honors came to him in his professional and civic life, and he also held many hospital appointments.

He seldom missed a meeting of the national ear, nose and throat societies, and in 1921 he was elected to the presidency of the American Otological Society. In 1934, he became president of the American Academy of Ophthalmology and Oto-Laryngology. The Edward J. Ill Award of the Academy of Medicine was conferred upon him in 1939, and in 1940 he was the recipient of the Award of Merit from his state medical society. The honorary degree of Doctor of Sciences was con-

ferred upon him by the University of Newark in 1942 and by Rutgers University in 1944.

In 1921, as chairman of the Welfare Committee of the New Jersey State Medical Society, he organized a thousand physicians for a demonstration for educational legislation.

Travel was his hobby and during many of his transatlantic trips he visited at the major ear, nose and throat hospitals throughout Europe.

As one of his colleagues writes, "he was a very modest individual with a profound knowledge and experience in otolaryngology and brain surgery." He was a constant student and a diligent worker. He will be remembered by all as a great educator and a pioneer in the development of his specialty.

Dr. Eagleton is survived by his widow, Mrs. Florence Peshine Eagleton.

CONTENTS